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ABSTRACT BOOKLET

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OC1

Assessing the feasibility of a digital behavioural intervention to support treatment adherence in young people (aged 13-17) with Inflammatory Bowel Disease

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Treatment non-adherence in adolescents with Inflammatory Bowel Disease (IBD) is as high as 93% [1]. Non-adherence to treatment has significant clinical consequences. Targeting this problem, we co-developed A Self-led Self-management Intervention Supporting Teens with IBD (ASSIST-IBD). ASSIST-IBD is a novel, user-centred and theory-driven digital intervention that supports young people (aged 13-17) with IBD to adhere to their treatment plan. Co-designed with adolescents who live with IBD, ASSIST-IBD consists of ten interactive modules aimed to increase young people's confidence to adhere to their treatment plan, support resilience to overcome adherence barriers, foster the development of health communication skills and generate optimism about the future. Within each module young people are supported to develop user-centred action plans to overcome their individual adherence challenges. These plans and corresponding behaviour change strategies are retained within the intervention modules, providing a personalised approach. A parent version of the intervention was also developed to support incremental transfer of responsibility for treatment adherence to young people themselves, as part of their transition to adulthood and adult services.

To assess the feasibility of ASSIST-IBD, young people with IBD (aged 13-17) who self-reported as being ≤80% adherent, and their parents, used the intervention for up to 12-weeks. Young people were able to complete modules in any order, based on their self-assessed adherence support needs. One-week post-intervention, young people and parents were invited to a qualitative interview to explore their experiences of using ASSIST-IBD, as well as their willingness to be randomised in a future Randomised Controlled Trial (RCT) and acceptability of more objective measures of treatment adherence (e.g., pill counts, biomarkers). Quantitative data was collected from the ASSIST-IBD digital platform on intervention engagement (e.g., number of logins, clicks on components, completeness of activities). Quantitative estimates of key parameters for a definitive RCT were also obtained (e.g., number of recruited participants; reasons for non-participation and ineligibility; retention and follow-up rates; reasons for early withdrawal; completeness and utility of outcome measures). Pre and post intervention measures examined changes in treatment adherence, quality of life and wellbeing. Two Public and Patient Involvements and Engagement groups of young people and parents co-analysed the study's data, allowing for their experiences and expertise to enrich the data analysis process.

Within post-intervention interviews, young people and parents reported a noticeable increase in young people's health self-efficacy, health communication and autonomous treatment adherence behaviours. ASSIST-IBD modules were generally rated favourably by young people, who believed the youth-led intervention content contributed to their engagement and behaviour change success.

The current research provides insight into the feasibility of ASSIST-IBD, and ways in which the intervention can be strengthened. This research is a valuable step in preparing for a definitive RCT to assess the effectiveness of ASSIST-IBD for young people with IBD.

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OC2

Evaluation of Vedolizumab Levels and response to treatment adjustments in paediatric patients with Inflammatory Bowel Disease (IBD)

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Vedolizumab is an anti-integrin antibody used in the treatment of paediatric inflammatory bowel disease (PIBD). Its use is recommended in individuals with moderately to severe active disease in which treatment with a tumour necrosis factor-alpha inhibitor (antiTNF-alpha) has failed, cannot be tolerated, or is contraindicated.[1]
Vedolizumab is safe and effective in PIBD. An exposure-response relationship is suggested with higher drug levels associated with improved clinical outcomes.[2,3,4] However, this has not been replicated consistently .[5,6] There is limited data on the monitoring and outcome of those on vedolizumab in contrast to those on antiTNF. Suggested levels are as follows: 37.1 mg/L at week 6, 18.4 mg /L at week 14, 12.7 mg/L during maintenance treatment.[7]

This study evaluates vedolizumab serum levels in PIBD patients, assessing the impact on subsequent treatment decisions and response.

Patients on vedolizumab were identified from the IBD Database. Data was collected on diagnosis, age at diagnosis, previous treatment (use of biologics, azathioprine, methotrexate, anti-TNF agents, and Ustekinumab), initiation date of vedolizumab, inflammatory markers, clinical symptoms, and endoscopic findings before and after treatment. Treatment adjustments were made based on vedolizumab levels and clinical symptoms.

23 children were identified, mean age at diagnosis was 9 years old (range 2- 15). 16/23 had Crohn's (luminal), 5/23 Crohn's with perianal disease and 2/23 Ulcerative colitis. 17/23 (73.96%) had initial treatment stepped up to biologics while 6/23 (26.09%) received biologics from diagnosis. 21/23 (91%) received antiTNF prior to switching to Vedolizumab. 2/23(8.7%) had received Ustekinumab. 19/23 (82.6%) were on Azathioprine and 10/23 (43.4%) were on Methotrexate.

21/23(91.3%) had levels taken. 2/23(8.7%) had recently started and not yet had levels. 6/21(28.6%) had levels taken during induction (8 – 14 weeks),15/21 (71.4%) in maintenance (after 14 weeks). Levels in induction were (mean:15.28mg/L, median:11.5mg/L , range:4.5-27.9). 2/5 (40%) met therapeutic levels.1 result was still in process at time of data collection. Levels in maintenance were (mean: 17.9mg/L, median: 8.2mg/L, range 3.5-80.9). 5/15(33.3 %) met therapeutic levels. 4/15 (26.7%) in maintenance had undetectable levels.

Treatment was changed based on levels in 11/21 (52.3 %), Changes done included adjusting frequency as well as dosing. In 5/21 (23.8%) vedolizumab levels > 12.7mg/L were associated with lower inflammatory markers. 5/23 had reassessment endoscopy. 3/5 showed improved endoscopic appearances with 2/3 of these having trough levels in range.

Most patients were escalated to vedolizumab following treatment with immunomodulator plus anti-TNF. While vedolizumab trough levels during induction and maintenance phase had a median of 11.5 mg/L and 8.2 mg/L respectively, no significant correlation was consistently observed between higher trough levels and reduced inflammatory markers. Standard dosing on induction and maintenance regimen was not sufficient to achieve target levels in 13/21 (61.9%). Adjustments in dosing as well as treatment intervals were made based on trough levels. Endoscopic improvement in 2/5 assessed correlated with trough levels in range, this supports a potential benefit of therapeutic drug monitoring in optimizing clinical outcomes. Further investigation is needed to determine the relationship between vedolizumab levels and clinical response in this population

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OC3

Very early onset IBD; monogenic causes and prognoses in a single tertiary centre study.

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Very early onset inflammatory bowel disease (veoIBD); defined as age at presentation of less than 6 years, accounts for up to 63% of monogenic IBD.[1] Over 80 monogenic causes have been identified; encapsulating primary immune deficiencies as well as epithelial defects.[2] The most recent European Society for Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) position paper on monogenic IBD recommends gene sequencing in all patients presenting at less than 2 years and any patients age 2-6 years with red flag criteria (family history, consanguinity, clinical features of immunodeficiency).[3] The advent of next generation gene sequencing technologies (such as targeted panel and exome/genome sequencing) means a wider range of variants can be tested with quicker turnaround times.[1,3]
Genetic panels were reviewed for IBD patients diagnosed aged 6 years or younger in a single tertiary paediatric centre. The aim was to quantify the proportion of the veoIBD cohort with a monogenic diagnosis, what these diagnoses were and effects on management and prognosis.

93 patients diagnosed with IBD at the age of 6 years or younger between 2010 and 2024 were identified from the departmental IBD database. Data was collected on gender, co-morbidities, family history, severity at presentation, genetic testing, management and clinical outcomes. Particular focus was then paid to any patients with pathogenic gene variants; analysing the changes made to their management and the patients' clinical remission status.

Diagnosis	Number of patients
Interleukin 10 deficiency	3
FMF	2
Chronic granulomatous disease	2
Cytoplasmic isoleucyl-tRNA synthetase (IARS) deficiency	1
Nucleotide-binding oligomerization domain-containing protein 2 (NOD2) defect	1
Tumour necrosis factor alpha induced protein 3 (TNFAIP3) related disease	1

Table 1: Summary of monogenic defects identified.

10/93 were aged under 2 years at presentation; all had genetics sent as per local guidelines. 20/83 (23%) patients aged 2 to 6 years at presentation had genetic panels sent as guided by red flag criteria and immunology review. 10/93 (10.7%) of the veoIBD cohort and 4/10 (40%) aged under 2 at diagnosis had a confirmed monogenic cause, in keeping with published rates of 0 -33% and 13-41% respectively.[3] 4/10 (40%) proceeded to bone marrow transplant (BMT), with 1 more planned for BMT this year. 2 patients received targeted treatment with colchicine for familial Mediterranean fever (FMF). 8/10 (80%) are currently in clinical remission and in 7/10 (70%) treatment for IBD has been discontinued.

Whilst monogenic IBD remains a severe disease entity associated with significant morbidity; it often has specific and effective treatment strategies.[3] Wider variant testing and quicker analysis mean these crucial diagnoses can be made earlier in presentation and therefore clinical remission can be attained in a group of patients once deemed treatment refractory.[1] Rates of monogenic disease were similar in this cohort to published data. In 7/10 identification of a monogenic cause has led to targeted treatment. Genetic sequencing should be considered in all patients presenting with veoIBD, those with red flags or those refractory to standard treatment.[3]

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OC4

Discontinuation of Premedication for Infliximab Infusions is Well-Tolerated, Without Adverse Events, and Cost-Saving in Paediatric Inflammatory Bowel Disease

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There is regional and national variability in the use of routine prophylactic medications for the prevention of anaphylactic or drug reaction with biologic therapies in inflammatory bowel disease (IBD) (1). Increasing reports have demonstrated the safety of IFX without premedication and that routine use of steroids and antihistamine medications may be unnecessary (2). The benefits of discontinuing routine premedication would be the reduction in medication side effects, cost and time taken for infusion visits. We aimed to demonstrate that the number of infusion reactions would not increase without premedication and to assess the response of children and young people with IBD and their families to this change.

Methods:
From the first of June 2024, premedication prescription was discontinued for patients attending the day unit at our specialist paediatric gastroenterology centre. We aimed to assess service user response to this change over two discrete two-month periods (June-July and August-September). These responses were collected through anonymised feedback forms. Participants were asked three questions in this way, with each question posing a "yes/no" response and allowing free-text space for elaboration on these responses. Questions assessed if the user had been informed of the change, if their infusion had gone differently to previously and whether they had experienced anything different over the course of days after the infusion. Over this time a record was made of all infusions given on the day unit and adverse events were recorded. Medication costs were calculated using the British national formulary for children (BNFc) (3).

Results:
A total of 175 infusions were recorded over the examined period and 35 questionnaires were completed. No reactions to infliximab were reported. Medication cost was calculated at an average value of £6.10 per visit; without nursing costs. This represents a saving of £1,067.5 over the four months examined.
Thirty of the thirty-five respondents reported having been informed of the change in practice (86%). Eighteen respondents noted a change in the experience of having the infusion (51%) though the changes were largely positive with participants reporting decreased lethargy or drowsiness. Fifteen respondents (43%) reported noticing changes after the infusion and in the subsequent days. Comments reported participants having "more energy," being "less unwell" and being "able to go to school" after the infusion. Some responded noting "less appetite" and a "sore arm". The most commonly used words were proportionally represented in Figure 1; a word cloud.

Conclusions:
Discontinuation of premedication for infliximab infusions was safe, cost effective and positive experience for our patients. Keeping patients informed of changes to their treatment is important to the team, which was demonstrated in responses. An immediate and subsequent reduction in tiredness is a positive outcome for this group who attend infusions around school, college and employment and could facilitate reduction in missed days. As well as reducing cost, the withdrawal of additional intravenous medications has reduced nursing time and overall infusion time.

Figure 1: A word cloud demonstrating the most used words and phrases in participant feedback.

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OC5

Refeeding syndrome in patients commencing exclusive enteral nutrition for paediatric Crohn's disease: a single centre experience

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Crohn's disease (CD) is an inflammatory disease of the gastrointestinal tract which can be associated with weight loss or faltering growth (1). When nutrition has been restricted the patient is at risk of developing refeeding syndrome (RFS) (2). The presence of hypophosphataemia is a key indicator of RFS. Other biochemical abnormalities can occur during the early stages of RFS, including low levels of potassium and magnesium. RFS can have life-threatening consequences (2)

Exclusive enteral nutrition (EEN) is advised by ECCO-ESPGHAN "in children with active luminal CD, dietary therapy with exclusive enteral nutrition [EEN] is recommended as first line for induction of remission" (1). With this increase in nutritional intake, patients may be at increased risk of RFS.

We aimed to evaluate whether children with newly diagnosed CD who underwent EEN as initial induction method were at risk of developing RFS, and if so if they would need treatment for electrolyte disturbance. We hypothesised that the risk of needing IV electrolyte replacement would be low.

60 patients were identified from the departmental IBD database who had been diagnosed with Crohn's disease over a 14 month period. Data was collected regarding demographics, anthropometry, EEN treatment, cardiovascular observations, potassium, phosphate and magnesium levels, and electrolyte replacement therapy. 7 patients were excluded as nutritional supplements had been initiated prior to EEN, 5 patients were excluded for insufficient biochemistry data. The remaining 48 patients were further divided into two risk groups based on percentage median Body Mass Index, weight loss and dietary intake.

In total, 19 patients (39.6%) were observed to have an electrolyte disturbance following initiation of EEN. Of these, 13 needed electrolyte replacement (27.1%) and no patients needed IV replacement.
There were 22 patients in the low risk group. Of these, 5 (22.7%) had electrolyte disturbances and only 1 (4.5%) required replacement which was oral.

Of the 26 patients in the high risk group, 14 (53.8%) had electrolyte disturbances, and 12 patients (46.2%) needed electrolyte replacement.
In 13 patients where electrolyte replacement was prescribed, all the patients needed oral phosphate replacement (100%) and 1 of these also needed oral magnesium replacement.

No patients were admitted to critical care for management.

This data suggests that the whilst the incidence of refeeding syndrome in this population is 39%, no patients required IV replacement despite their initial refeeding risk group. All patients were managed with oral supplements. Fewer patients identified as low risk required replacement. Patients are often admitted to start EEN due to concerns over refeeding syndrome but this study suggests community based EEN with blood monitoring, dietetic input and clinician overnight may be safe from a refeeding perspective.

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OC6

Evaluation of the use of Jorveza (orodispersible budesonide) for eosinophilic oesophagitis in a single tertiary paediatric centre.

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Eosinophilic oesophagitis (EOE) is an antigen mediated inflammatory disorder of the oesophagus; defined by symptoms of oesophageal dysfunction and histological findings of eosinophil infiltration with an eosinophil count of more than 15 per high power field.[1] Management strategies include dietary eliminations, proton pump inhibitor (PPI) therapy and topical steroids. Oral viscous budesonide (OVB) is most commonly used but has limitations in difficulty of making up the slurry, difficulty obtaining the sweetener used to mix the solution, and parental concern about use of artificial sweetener.[2] Jorveza (orodispersible budesonide) is an alternative preparation but is only licensed in over 18 years.[2]

The aim of this study was to evaluate management strategies in patients with EOE in a single tertiary paediatric centre. Specifically looking to compare the use of Jorveza in the paediatric cohort compared with more traditional treatment strategies (PPI, dietary exclusion, OVB or combination therapy). The hypothesis was that better clinical and histological remission rates would be seen in the Jorveza cohort.

A retrospective review of patient records identified 45 patients diagnosed with EOE between 2012 and 2024 (41 male, 4 female) with age ranging from 2 to 16 years at presentation. Data was collected on symptoms at presentation, histological findings, therapeutic management and clinical remission status.

13 patients were treated with monotherapy (either PPI, Jorveza, OVB or dietary exclusion) and 32 received combination therapy (any combination of the above). 9 patients in total were treated with dietary exclusion- 1 with exclusion alone; 8 with combination medical and dietary therapy. 1 patient had a six food elimination diet and 8 eliminated 1-2 food groups, most commonly dairy. Of the 9 patients who received Jorveza/PPI 6 (85%) were in clinical remission (the remaining patient had not yet been reassessed). The 2 patients receiving Jorveza monotherapy are awaiting reassessment. 13/17 (76%) patients who received OVB/PPI were in clinical remission. 2/4 (50%) patients who were treated with PPI/OVB/dietary exclusion were in clinical remission.

The numbers remain small; but this study reflects slightly improved clinical remission rates in patients using Jorveza. This may be explained by ease of administration, better tolerance and therefore improved compliance compared to OVB in paediatric patients. This study also highlights the inconsistent implementation of elimination diets based on patient reported symptoms and specific IgE allergy testing. Ease of access to endoscopy is imperative in making clear decisions about elimination strategies. Close working with the dietetic department to formulate a pathway for dietary exclusions is vital. With a multitude of potential therapeutic combinations available, a stepwise approach catered for the patient and their family should always be utilised.[1,3] Jorveza seems to be a promising option for patients with EOE but more data needs to be collected on both clinical and histological outcome.

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OC7

The prevalence and clinical significance of Donor Specific Antibodies in paediatric liver transplantation

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The role of donor specific antibodies (DSA) in allograft injury is poorly understood in liver transplants (LTx)[1]. DSA have potential associations with ductopenia, biliary strictures and accelerated fibrosis [2].

A retrospective analysis was performed to understand the prevalence of DSA in LTx children attending protocol liver biopsy (LB) and correlate the association of DSAs with histopathological and biochemical findings [4].

Demographic details of LTx children undergoing simultaneous protocol LB and DSA testing from Jan 20- Dec 23 were included 115 children (age -15 months to 18 years, median – 11 years, 63 male and 52 female) had DSA testing performed around the time of liver biopsy. 41/115 patients (36%) had positive DSAs.

37/41 children (90.2%) had DSA against HLA class II, HLA-DQ being the most prevalent (70.3%). 35 patients (73%) had mean fluorescence intensity (MFI) levels > 10,000 (5 patients had acute cellular rejection and 2 had chronic antibody mediated rejection).

Children with negative DSA had statistically significant normal liver biopsy (p 0.016) compared to DSA positive group. Acute cellular rejection (ACR) was seen in both DSA positive and DSA negative children with no statistical significance (p 0.098). Children with Positive DSA were more likely to have moderate -severe fibrosis (n=7, p 0.04). Their prior protocol biopsies showed no significant changes (n=4) or mild fibrosis (n=3) with negative DSA.

Three patients with chronic antibody mediated rejection (AMR) had de novo DSA against HLA-DQ and/or -DQA and their protocol biopsy post liver transplant was > 5 years. All of their previous protocol biopsy showed ACR with negative DSA. These patients were on dual or triple immunosuppression at the time of biopsy with normal liver biochemistry.

There was no significant difference in liver function test and immunosuppression (single vs dual/triple and tacrolimus levels) between DSA positive and negative groups. There was no correlation of ACR or graft fibrosis with MFI titres and hence have to be used in conjunction with liver

ACR was seen in post LTx children with DSA positive as well as DSA negative children, thus confirming uncertainty about the role of DSA in contributing to ACR. There should be a high index of suspicion in children who change status from DSA negative to DSA positive for detection of children evolving into AMR. The presence of DSA with high MFI titres on its own without a liver biopsy do not give any further information about the graft function or abnormalities

DSA in combination with protocol biopsies may help in understanding the underlying pathogenetic mechanisms contributing towards graft fibrosis, which may further improve long term outcome in liver transplantation.

Table: Findings on protocol liver biopsy

Histopathology	DSA negative (n=74)	DSA positive (n=41)	p value
No significant changes	22 (29.7%)	3 (7.3%)	0.016
Acute cellular rejection	9 (12.1%)	9 (22%)	0.098
Acute antibody mediated rejection	0	0	
Chronic antibody mediated rejection	0	3 (7.3%)	0.018
Mild fibrosis	37 (50%)	18 (43.9%)	0.57
Moderate to severe fibrosis	4 (5.4%)	7 (17%)	0.042
Cirrhosis	0	0	

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OC8

Tracing the timeline: Is Fontan-associated liver disease a paediatric concern? A retrospective longitudinal study

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The Fontan procedure is the definitive surgical approach for managing congenital heart defects causing functionally univentricular physiology[1]. Performed between 4-6 years of age, it involves connecting the systemic venous return to the pulmonary circulation. However, over time, the Fontan circulation causes venous congestion and decreased cardiac output, leading to hepatic fibrosis and ischemia[2]. Consequently, patients develop Fontan-Associated Liver Disease (FALD); the most common cause of end-organ dysfunction in Fontan patients[3]. Although this phenomenon is well-documented within the literature, no study clarifies when FALD begins. This study aimed to determine the initiation and extent of FALD in the paediatric population, thus helping inform effective follow-up protocols for liver surveillance post-Fontan.

We conducted a retrospective longitudinal analysis of 353 Fontan patients at a major paediatric centre in England. Patients born between 2003-2022 who underwent a Fontan operation before 2023 were included. Patients who subsequently underwent heart/liver transplants or were lost to follow-up were excluded. LFTs and FBCs were obtained for each patient preoperatively and postoperatively at 5,10, and 15 years, where available. The variables included Bilirubin, ALP, ALT, AST, GGT, Albumin and Platelets. At each time point, an abnormal LFT was defined as a derangement of any of the above variables, and these results were compared between time points. Further comparison of levels of each variable between time points was performed using the Wilcoxon signed-rank test. The extent of liver damage was also assessed using post-Fontan ultrasound scans (USS).

Preoperative, 5-year, 10-year, and 15-year postoperative LFTs were available in 155/353, 298/353, 166/353 and 51/353 patients, respectively. Of these, the proportions of abnormal LFTs were 17%, 56%, 71% and 63%, respectively. Table 1 summarises the statistically significant changes in each variable between the preoperative and postoperative time points. No analysis was conducted for AST and GGT due to missing data. Pertinently, Bilirubin ($p=0.012$), ALT ($p<0.001$) and Platelets ($p<0.001$) levels showed significant changes within the first 5 years post-Fontan. Changes in Albumin and ALP were attributed to confounding variables, including bone growth and Fontan comorbidities. 82 USS were available at a median of 8 [IQR: 5-10] years post-Fontan, with 59% exhibiting abnormal echotexture. In three cases, focal lesions were visualised on USS but were confirmed to be normal on biopsy and MRI.

Although it is uncertain whether the statistically significant LFT changes correlate to clinical significance, we can conclude that the liver experiences some insult within the first 5 years post-Fontan. In current practice, global follow-up protocols for FALD surveillance begin 5-10 years post-Fontan. Considering our findings, we propose preoperative and annual postoperative LFTs be performed for all Fontan patients. Furthermore, we recommend that paediatric centres introduce and regularly collect a Fontan liver panel for all patients to enhance early detection of FALD. These may be reviewed periodically in Fontan liver clinics where Cardiologists and Hepatologists work in conjunction, as has been successfully implemented in our centre.

Table 1 – Summary of LFT changes post-Fontan

	Pre-op vs 5-year	5-year vs 10-year	10-year vs 15-year
Bilirubin	↑	↑	↔
AST			
ALT	↑	↔	↔
ALP	↓	↑	↓
GGT			
Albumin	↔	↑	↔
Platelets	↓	↓	↔

Table showing LFT changes between time points. ↑ = significant increase. ↓ = significant decrease. ↔ = no significant change.

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OC9

Closure of hepatic congenital portosystemic shunts using cardiac closure devices: a case series

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This report details the first UK-based experience of closing congenital portosystemic shunts (CPSS) using intravascularly deployed cardiac closure devices. The procedure, performed by interventional radiologists at a single specialist centre, involved five paediatric patients between January 2019 and September 2024.

CPSS are rare vascular malformations resulting in the bypass of hepatic sinusoidal blood flow.[1,2] They can be classified as being extra- or intra-hepatic, and further described by the connecting vessels.[1,2] Complications are secondary to physiological compounds evading metabolism within the liver.[3,4] This can result in significant morbidity including varying degrees of neurocognitive impairment, hepato-pulmonary syndrome, pulmonary hypertension, liver tumours and endocrine abnormalities.[3,4]

Approaches to closure were traditionally surgical and have progressed to less invasive methods with interventional radiology avoiding morbidity associated with laparotomy.[2]

In selected cases, CPSS were deemed unsuitable for endovascular closure with standard available vascular devices. Following careful multidisciplinary team discussion, we considered the use of cardiac septal occluder devices that would best suit the anatomy of the CPSS and avoid laparotomy. We made an application for a new interventional procedure for off-label use of cardiac devices in these patients. Patient families were appropriately consented.

Four of the patients received an atrial septal defect occluder and one received a ventricular septal defect occluder. The devices ranged from 9mm to 27mm in diameter.

Table summarising patient data:

Age at diagnosis (median)	4 years 6 months
Age at device use (median)	13 years 4 months
Time between diagnosis and closure (median)	6 years 3 months
Clinical presentation of CPSS	3 were incidentally diagnosed 1 had conjugated hyperbilirubinaemia 1 had a liver tumour
Number of shunts	All had 1 shunt
Shunt anatomy	4 had main portal vein to inferior vena cava (side to side connection) 1 had main portal vein to right atrium
Liver nodules	4 of 5 had liver nodules (1-5 in number)
Cardiac comorbidities	Atrial septal defect, cardiomegaly, patent ductus arteriosus, enlarged superior vena cava, mild aortic stenosis and tiny patent foramen ovale
Other comorbidities	Failure to thrive, poor concentration, attention deficit hyperactivity disorder, developmental delay, premature adrenarche, polycystic ovarian syndrome, hyperandrogenism and insulin resistance
Single stage closure	3 of 5
Anticoagulation	All patients received subcutaneous heparin post operatively 1 case that suffered thrombosis received warfarin for 2 years 3 cases stopped heparin by 1 year of follow up
Complications	1 patient had device migration, twice 1 had thrombosis
Outcome	1 patient remains untreated (device migration) 4 are off anticoagulation and well.

These cases highlight the feasibility of using cardiac closure devices in CPSS with anatomy that may seem unsuitable for endovascular closure. Although there have been challenges in initiating a new technique, overall results have been promising and are improving with increased experience.

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OC10

Analgesia in acute paediatric pancreatitis

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Acute pancreatitis (AP) in children and young people (CYP) is a rare disease occurring in less than 1 per 100,000. This presents challenges in ensuring an accurate diagnosis of AP is made and considered. Responding to pain in a timely manner and monitoring pain levels remains a challenge in the paediatric population. We evaluated the pain management of patients presenting with AP to our tertiary children's hospital.

Elevated lipase results were identified through our trusts biochemistry laboratory between September 2022 to June 2024 in CYP less than 18 years old. Electronic patient records were reviewed, and patient-episodes identified that meet NASPGHAN[1] diagnostic criteria for AP (2 or more: abdominal pain consistent with AP, serum lipase levels 3 times greater than the upper limit of normal, or imaging consistent with AP). Episodes of AP were excluded if notes were incomplete, managed by adult gastroenterology, died due to other medical causes or remains a current inpatient.

967 episodes of elevated lipase levels were identified. 60 episodes met criteria for inclusion. Age ranged 2–16 years [average 12 years]. Diagnoses: idiopathic [13], mumps [1], gallstone (GS) pancreatitis [13], acute recurrent pancreatitis (ARP) [11], SPINK1 mutation [1], PRSS1 mutation [1], propionic academic (PA) [5], methylmalonic academia (MMA) [5], azathioprine-induced [3], chemotherapy-induced (PEG-asparaginase) [14], post-operative complication [2] and post-ERCP [1]. 7 patients required an inpatient cholecystectomy, and 2 patient-episodes of GS pancreatitis required inpatient ERCP.

Imaging included abdominal US [51], CT [18] and MRCP [20]. 17 patient-episodes were NBM, 37 received intravenous fluids and 15 received parenteral nutrition. 30 patient-episodes received intravenous antibiotics.

17 patient-episodes were managed with enteral analgesia (paracetamol and oramorph/oxycodone) with an average length of stay of 9 days [1–51 days]. 18 patient-episodes escalated to IV analgesia with an average length of stay of 18 days [2–100 days]. 25 patient-episodes required escalation to opioid infusions with an average length of stay of 28 days [3–239 days]; chemotherapy-induced [8], MMA [1], PA [3], GS-pancreatitis [7], idiopathic [1], post-operative complication [1], ARP [3] and SPINK1 [1]. 5 patient-episodes required paediatric critical care (PCC) for pain management: GS-pancreatitis [2], chemotherapy-induced [1], PA [1] and idiopathic [1].

The average length of stay was 9 days [1–239 days]. Patients that underwent cholecystectomy [7] remained an inpatient for an average of 12 days [5–51 days]. Patient-episodes with a length of stay >10 days included: idiopathic [3], mumps [1], GS-pancreatitis [6], post-operative complication [2], ARP [2], PA [2] and chemotherapy-induced [10]. Complications included 4 patient-episodes of necrotising pancreatitis; idiopathic [2], GS-pancreatitis [1] and chemotherapy-induced [1]. 3 required escalation to PCC.

Active management of pain is important in the patient journey. 42% of our patient-episodes required escalation of analgesia to opioid infusions suggesting that earlier intervention with specialist support from the acute pain service will improve pain management and potentially shorten the length of stay. Following this review, we are creating a flow chart to streamline analgesia escalation and provide education and training to members of staff looking after CYP with AP.

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OC11

Vascular complications following liver transplantation- a single Centre experience over 10 years

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The long-term outcome of pediatric liver transplantation (LTx) is determined by complications(vascular, hematological and infectious etc) that occur both immediately post-transplantation and in the long-term***(1).*** We aimed to analyze the pattern and risk factors for vascular complications (VC) in a single tertiary LTx center. A retrospective analysis on children undergoing LTx over 10 yrs (2013- 2023). Liver unit database, medical records were reviewed in different time periods post-transplant (week, 3m , 6m , 1 year, 5 yr, 10 yr, 15 yr for vascular complications e.g hepatic artery thrombosis (HAT) and stenosis (HAS) , portal vein thrombosis (PVT) and stenosis (PVS) and Hepatic vein thrombosis (HVT).

table 1 :Demographic and vascular complications: n=300

Age median (IQR in years)		7.00 (3.00–11.00)
Weight median (IQR in kg)		15.00 (8.80–29.00)
Diagnosis	Biliary Atresia	103 (34.3%)
	Acute liver failure	38 (12.7%)
	Autoimmune liver disease	19 (6.3 %)
	Metabolic liver disease	69 (23%)
	Hepatoblastoma	40 (13.3%)
	Others	31 (10.3%)
Height median (IQR in cm)		92.20 (73.50–130.70)
Early Hepatic Artery Thrombosis with in the first month		12 (4.0%)
Late Hepatic Artery Thrombosis after the first month		8 (2.7%)
Hepatic artery stenosis		6 (2.0%)
Portal vein thrombosis		17 (5.6%)
Portal vein stenosis		15 (5.0%)
Hepatic vein thrombosis		3 (1.0%)

In early HAT : 2/12 underwent re-transplantation and 10/12 children underwent surgical treatment (thrombectomy n=6 , re-anastomosis n=3 , thrombectomy and re-anastomosis n=1). 7 /20 with HAT died due to multi organ failure, bleeding or biliary sepsis (median time to death 9 days ,range from 1 -790 days) Multivariate analysis found significant correlation between incidence of HAT with split graft, cold ischemia time, recipient weight less than 10 kg.

HAS 6 patients post-transplant: 2/6 had balloon angioplasty,3/6 of them had stenosis of only one segment of HA, with patient main HA, normal LFTS and were maintained on anticoagulation, 1 died due to MOF and sepsis

PVT occurred in 17 patients (median 4 days, range 1-1155 days) 9/17 had early PVT (n=5 re-exploration and patency, 3 non-occlusive thrombus- conservative management, 1 had Interventional radiology). All of them survived immediate intervention and in late PVT 8/17- 3 died median time to death is 545 days (range 90 – 1245 days). Multivariate analysis found a significant correlation between recipient weight less than 10 kg, donor-recipient size mismatch, split liver graft and pre-existing portal vein condition.

Biliary atresia was a common diagnosis in children with HAT 6/20 (30%) and PVT 9/17 (45%) . The other common condition in HAT was metabolic liver disease.3 patients developed HVT (median range) and had stents inserted by hepatic venography and were maintained on long term anticoagulation.

Recipients with less than 10 kg, split graft recipients and longer cold ischemic times had a higher incidence of vascular complications. A high index of suspicion needs to be maintained in the evaluation of the children with biliary atresia and metabolic liver disease patients peri-operatively and in long term follow-up.

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OC12

EndoFLIP assessment of oesophageal sphincter in children: a single-centre experience

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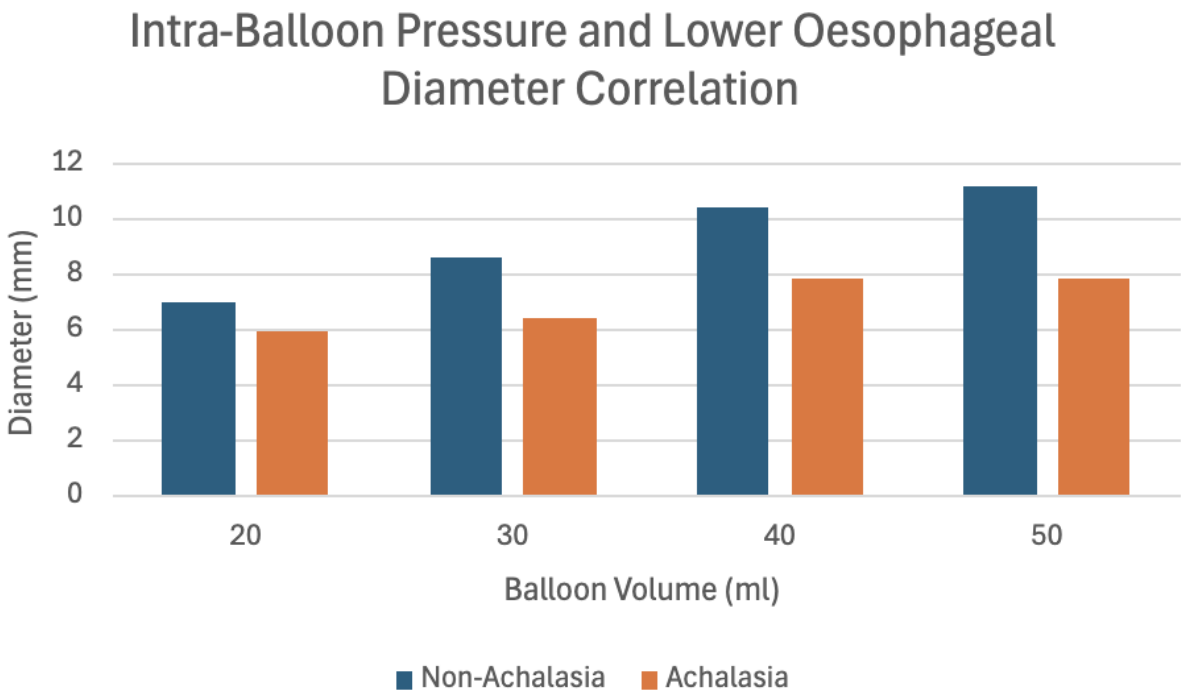
Assessing the oesophageal sphincter in children is vital for diagnosing gastrointestinal disorders such as gastroesophageal reflux disease (GORD) and achalasia. However, normative data for paediatric oesophageal function are limited, hindering clinical interpretation. EndoFLIP (Endoluminal Functional Imaging Probe) provides a dynamic evaluation of the sphincter by measuring diameter, compliance, pressure, and distensibility. Despite its utility in adults, baseline oesophageal values in children remain largely unexplored. This study aims to establish baseline oesophageal sphincter values in a paediatric population using EndoFLIP differentiating between those with achalasia and those without, to enhance diagnostic accuracy, providing reference data to facilitate the evaluation of oesophageal function in clinical practice

This retrospective review was conducted at our single-centre institution, including paediatric patients aged 1 to 16 years who underwent EndoFLIP assessment between June 2022 and May 2024. The EndoFLIP was performed during endoscopy, comparing data between achalasia and non-achalasia groups.

A total of 274 measurements were collected from 32 children, mean age 9.5 ± 4.6 years; 53% male, showing distinct trends in key parameters as the balloon volume increased. Data were grouped into achalasia (n=6) and non-achalasia (n=26) cohorts. Measurements (non-achalasia and achalasia) included oesophageal diameter, compliance, pressure, and distensibility, categorised by balloon volumes of 20, 30, 40, and 50 mL. Diameter (7.0, 5.9), (8.6, 6.4), (10.4, 7.8), (11.2, 7.8) mm. Compliance (116.6, 87.8), (127.3, 189.4), (117.0, 194.0), (179.2, 205.0) mm^3/mmHg. Pressure (14.1, 10.7), (22.9, 12.7), (29.3, 18.5), (36.8, 23.8) mmHg. Distensibility (3.1, 3.1), (3.1, 3.1), (3.6, 2.9), (3.4, 2.2) mm^2/mmHg.



Overall, these results reveal clear differences in oesophageal diameter, compliance, pressure, and distensibility between groups across increasing balloon volumes, providing key reference points that distinguish achalasia from non-achalasia presentations, establishing normative values for oesophageal function in children and enhancing understanding of oesophageal motility in this population. Achalasia patients had smaller oesophageal diameters, lower initial compliance, lower resting pressures, and decreasing distensibility with balloon volume compared to non-achalasia patients. Non-achalasia patients showed increasing diameter and stable distensibility with volume, while achalasia patients exhibited reduced compliance, characteristic of achalasia. This study establishes normative EndoFLIP values for the oesophageal sphincter in a paediatric population, offering a valuable reference for assessing oesophageal function in children, using balloon volume less than 40ml during EndoFLIP does not allow accurate evaluation of LOS. The findings demonstrate clear trends in diameter, compliance, pressure, and distensibility across increasing balloon volumes with balloon volume 40 and 50ml are accurate at assessing LOS disorders, enhancing our understanding of oesophageal motility and aiding in the evaluation of paediatric gastroenterological conditions.

OC13

Comparison of quality of life in paediatric functional constipation in different age groups: A case-control study

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Functional constipation (FC) in the paediatric age group is a significant global health issue, with prevalence rates varying across geographic regions. In the UK, it is estimated at 10-20%. FC accounts for a notable percentage of paediatric consultations and referrals. The condition negatively impacts children's physical, emotional, social, and educational well-being and poses a burden on families and healthcare systems. Although much is reported on various metrics related to childhood constipation, there is a paucity of data on the impact of this chronic and common condition on the quality of life of young people. Measuring quality of life is vitally important in shaping services to meet community needs. This study aims to report the impact on the quality of life of children and young people in the XX Network using a validated health-related quality of life tool, PedsQL.

Aim is to compare the quality of life (QoL) between children with functional constipation and their healthy peers using the Paediatric Quality of Life Inventory (PedsQL).

This case-control study was conducted with 112 children being treated for constipation and 35 healthy controls, aged 2-16 years. The study spanned multiple centres across XX Region, UK, from October 2021 to November 2023. The PedsQL, a validated questionnaire covering physical, emotional, social, and school functioning domains, was used to assess QoL. Data were analysed using the Mann-Whitney U test, with p-values <0.05 considered statistically significant.

Children with constipation had significantly lower QoL scores across all domains compared to healthy controls. Physical functioning scores were 60.00 ± 24.43 (constipation) vs. 86.60 ± 13.40 (control) ($p<0.001$). Emotional functioning scores were 47.65 ± 21.94 (constipation) vs. 76.85 ± 19.40 (control) ($p<0.001$). Social functioning scores were 64.57 ± 26.73 (constipation) vs. 85.00 ± 20.10 (control) ($p<0.001$). Educational functioning scores were 59.71 ± 26.34 (constipation) vs. 79.23 ± 20.92 (control) ($p<0.001$). Parents reported lower scores compared to children, particularly in emotional functioning ($p=0.003$).

Children with functional constipation report a significantly lower quality of life compared to their non-constipated peers, affecting multiple aspects of daily functioning. The chronic nature and broad-reaching impacts of FC highlight the need for improved management strategies to enhance QoL for affected children and their families. Further research is needed to explore the impact of treatment on QoL in paediatric populations.

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OC14

"My child doesn't know when they need a poo!"- Parent and Patient Reported Symptoms do not Predict Rectal Sensation.

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Background
The prevalence of constipation in children, affects between 0.7 and 29.6% of the general population in UK and worldwide (1,2), and is frequently associated with faecal incontinence (3). The coexistence of functional constipation and faecal (FCFI) has been recognised within paediatrics (4).

Aim: We aim to compare subjective reporting using bowel questionnaire with objective findings (using high resolution anorectal manometry - HRAM).

Methods
A prospective maintained database was reviewed for patients in the service between September 2016- October 2024. Data regarding demographics, questionnaire (St Mark's Incontinence Score - SMIS) and HRAM. SPSS was used to perform statistical analysis: median, range, percentages, and Pearson correlation (p value of <0.05 as significant).

Results
In total, 288 patients completed the SMIS questionnaire and underwent awake HRAM. 58% males; median age 10-year-old (range 1- 27 years); 27% with neurodiversity. There was no correlation between parents reporting and physiological findings ($p>0.05$).

Table 1: Compare subjective and objective findings.

		Subjective reported symptoms (SMIS)	
		Normal (SMIS)	**Abnormal (SMIS)**
Objective measurements (HRAM)	Normal Sensation	54 (19%)	162(56%)
	Abnormal Sensation	17 (6%)	55 (19%)

Conclusion
1. Subjective findings do not correlate with objective HRAM findings.
2. There is a gap between parental reporting of symptoms and objective HRAM findings.
3. This gap could be explained by conscious denial (withholding), subconscious denial , impaired sensory processing (interoception), imperfect physiological testing and imperfect parental interpretation.

This finding demonstrates the importance of objective testing for a condition that relies heavily on parental reporting of the patient's symptom.

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OC15

Delivering a workshop to empower young people and their parents to improve understanding and management of the gut-brain interaction

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Disorders of the gut-brain interaction (DGBI) occur due to a bidirectional communication impairment between the gut and the brain. DGBI's are common in paediatric populations and can result in difficult clinical experiences for children and young people due to the complexities associated with diagnosis and management. The mechanisms which result in a DGBI diagnosis are unfortunately not routinely well defined, which can result in frustration and confusion for children and families. Moreover, DGBIs can be associated with significant functional impairment, decreased physical inactivity, reduced school attendance, and result in lower quality of life.

Given these challenges, the Paediatric Gastroenterology Psychology Service created and delivered a workshop to support young people managing their DGBI. The one-day workshop was based on principles from Cognitive Behavioural Therapy (CBT) and Acceptance and Commitment Therapy (ACT) to support their understanding of DGBI as well as empower them to live well alongside their symptoms. To consolidate learning, attendees were also given workbooks with the workshop content and space to complete exercises. Additionally, there was also a follow up webinar which informed parents of the skills and strategies discussed to support their child to apply the psychological interventions.

Three workshops ran from August to October 2024, with 16 participants aged 8 - 17 split into either the primary aged workshop or senior school aged workshop, respectively. All attendees completed feedback on the workshop using a Likert rating scale alongside free-text comments which were analysed using content analysis. Feedback forms were adjusted to be age appropriate. Prior to the workshop and at one-month follow up, attendees were asked to complete the Wellbeing and Health Experiences Evaluation Log (WHEEL).

On average, the senior school attendees (n=11) rated finding it useful to find out more about the gut brain interaction as 8/10; learning about stress and strategies to manage as 8.8/10; the workshop helped them to understand more about their symptoms and how to manage as 8.6/10; enjoyment of the workshop at 8.5/10; and likelihood of recommending the workshop to other young people with a similar diagnosis as 8.5/10 (where 1 is 'not at all' and 10 is 'very much'). Similar patterns of agreement were seen across the primary school attendees (n=4).

Themes from the qualitative feedback demonstrated the importance young people place on connecting with other young people who have similar DGBI experiences: *'I have learnt I'm not alone'.* They described the benefit of learning strategies they can apply to their symptoms: *'The CBT formulation helped me understand myself'.* This resulted in them taking away messages of validation and empowerment: '*I have learned that my feelings and experiences because of DGBI are valid so I can be kinder to myself'*.

Following the workshop, young people and their parents reported increased understanding of the gut brain relationship and found it helpful to understand various strategies to manage their symptoms. In addition, meeting other young people with similar presentations reduced feelings of isolation, which warrants further investigation on the impact on feelings of shame and stigma around gastroenterology symptoms.

OC16

Growth and long-term outcomes of children attending specialist multi-disciplinary cardiac feeding clinic at a tertiary centre between 2015-2023.

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Congenital heart disease is the most prevalent birth defect in the UK (1) significantly impacting growth and nutritional status in affected children (1-3). The Cardiac Multi-Disciplinary Team (MDT) Feeding clinic comprises gastroenterology, Speech and Language Therapy (SALT) and dieticians. It employs a multidisciplinary approach to enhance oral feeding, growth and nutrition in children with congenital cardiac disease.

The aim of this study was to assess outcomes of children attending the Cardiac MDT feeding clinic between 2015 and 2023 with focus on oral feeding status and growth.

A retrospective study was conducted, analysing data of the 200 children attending this clinic from 2015-2023. Patients were identified from hospital record systems with records reviewed for PEG status, feeding regimens (including oral intake), medications and growth measurements using WHO growth charts. Centile trajectories for height and weight were calculated.

145 (73 %) required PEG or PEG-J feeding, mostly due to long-term NG tube dependence (66%). Other reasons included feeding difficulties (20%) and unsafe swallow (19%). The median age of PEG insertion was 7.6 months (range 0.25-3.69). The median age for the first clinic was 7.9 months (range 0.10-7.16). 10 children (5%) died. 53 (27%) had a sole cardiac diagnosis.

Table 1: Comorbidities of patients attending the clinic

Co-morbidity	Number of patients (N)
Neurological	22 (11%)
Surgical/Anatomical	60 (30%)
Behavioural	15 (8%)
Genetic/Chromosomal	77 (39%)
Other	52 (26%)

122 (61%) children were prescribed anti-reflux medication at first clinic attendance compared with 75 children (38%) on their last. The most prescribed anti-reflux medication was omeprazole with 72 children (59% of those prescribed anti-reflux medication) prescribed this at first clinic attendance and 50 (67%) at the last.
At last follow-up, 94 (65%) patients had a PEG with 76 (81%) using it for feeding, and 14 (10%) patients had a PEG-J. 4 (19%) with a PEG used it for medication only or did not use the PEG. 109 (75%) were able to establish some level of oral feeding with 17 (39%) of these previously PEG-fed patients achieving oral feeding exclusively. 27 (14%) children with a PEG in situ used sole PEG or PEG-J feeds.
120 children (60%) showed an increase in centile change for weight and 100 (50%) for length. Only 21(11%) children recorded a decrease in weight centile and 28 (14%) height. All children with a decrease in weight had a PEG placed at some point as did 26 (93%) of those with a drop in height centile, with 5 (10%) of children in this group dying during the study. There was no gender difference in centile change during this time ($p>0.1$).

Most children previously PEG-fed were established on some level of oral feeding, and, in a significant number of patients, their growth was optimised too. Limitations exist due to those lost to follow-up. Centile data is also influenced by other factors, including other co-morbidities(4, 5). Further research to assess the impact of improvement in cardiac status and how this relates to success in oral feeding in this patient cohort would be beneficial.

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OC17

Nutritional blood monitoring (NTBM) demonstrates excellent safety profile for blended diet fed children – Recommendations for practice

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The administration of blended diet (BD), instead of commercial enteral formula was, until recently, considered out-with standard safe practice.[1] BD is increasingly considered as an alternative strategy for feeding in patients with Gastrointestinal Dystonia (GID). There is limited data on NTBM in the long-term BD population. We aimed to retrospectively review NTBM for BD within our regional service and make recommendations for practice from this data.

Patients commenced on BD between 1/1/17-1/5/24 were identified from five regional dietetic databases. Baseline demographics, diagnoses, patient complexity and the presence of GID were gathered. Patients were established on BD by local process which includes; dietetic assessment of nutritional requirements, risk assessment and an educational package. BD volumes were prescribed in combination with enteral feeds and titrated upwards as tolerated; individualised dietetic plans were reviewed for nutritional adequacy and adjusted via MDT forum. BD patients were defined as either full or partial BD (commercial enteral formula used in combination with BD). Local guidance recommends NTBM at baseline,6 months, and annually thereafter for; U+E, LFT, Bone Profile, PTH, Vitamins A, B12, D, E, zinc, copper, selenium, magnesium, FBC, ferritin and folate.[2] Patients were screened for inflammatory response using serum albumin and C-reactive protein.

85 patients were established on BD. 25 (29%) of these patients had GID. 23 (27%) patients received concurrent vitamin supplementation. 65 patients received partial BD. 24 (28%) patients received full BD. 5 patients had NTBM as per guidelines, 44 patients had an initial set at 6months, and 26 patients had an initial set 1 year after commencing BD. Abnormalities in NTBM identified ≥6 months after commencing BD are outlined in table 1.

Table 1

Blood Indices (Number monitored)	**Abnormality (number of patients)**	**Action (number of patients)**
Adjusted calcium (52)	Mildly elevated (3) Mildly deficient (1)	Alfacalcidol stopped (1) None (2) Dietary modification (1)
Copper (43)	Mild elevation (3) Mild deficiency (3)	Monitored (3) - no action Supplementation (1) None (2)
Ferritin (48)	Moderate-Severe deficiency (17)	Dietary modification (2) Supplementation (15)
Folate (37)	Mild deficiency (2)	Supplementation (2)
Magnesium (34)	Hypo/Hypermagnesemia (0)	
Plasma Selenium (42)	Mild elevation (3) Mild deficiency (6)	Not required - Red cell selenium normal (9)
Vitamin A (42)	Mild elevation (11)	Dietary modification (2) None (9)
Vitamin B12 (45)	Mild elevation (12)	None (12)
Vitamin D (52)	Insufficient (8) Mild deficiency (1)	Supplementation (6) None (3)
Vitamin E (39)	Mild elevation (23)	None (3) Not required - Vitamin E: cholesterol ratio normal (20)
Zinc (38)	Mild-moderate deficiency (11)	Supplementation (2)

The frequency and severity of NTBM abnormalities compare favourably with previous review of enteral or orally fed patients with neuro-disability.[3] We postulate that these favourable outcomes with NTBM reflect careful patient selection and intensive dietetic support. Under this pathway, we postulate that routine NTBM for Vitamin E, Copper and Selenium are not required in BD, and that NTBM is required only annually if first NTBM are within normal limits. Whether adjuvant enteral formula alongside BD diet helps to normalise micronutrients, or if vitamin supplementation is required warrants further research.

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OC18

Nutrition service 10-year review: impact on parenteral nutrition usage and wastage

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At a specialist paediatric centre, with 378 beds including a 31-bed paediatric intensive care unit, an expert Nutrition Support and Intestinal Failure Team (NSIFT) was established in 2014. This funded team was designed to oversee parenteral nutrition (PN) provision for all type 1 and 2 intestinal failure patients, with an aim to reduce PN wastage and increase PN standardisation. NSIFT review approximately 400-450 patients per year. The hospital does not have an aseptic manufacturing unit, therefore all PN is procured from external suppliers with strict deadlines. PN is provided either as bespoke/compounded bags or standard bags.

The aim of this project was to review if NSIFT have had a sustained impact on PN usage (bespoke versus standard) and PN wastage over the last 10 years.

Data was collected from the pharmacy PN database from 2016 onwards, due to data record availability):

- Number of PN referrals
- Bespoke versus standard PN numbers and costs
- Percentage of wasted PN

NSIFT activity
Average number of PN referrals was:
In 2016, average 26/month
In 2020, average 39/month
In 2024, average 32/month (23% increase in PN referrals)

PN usage
Total PN provision (mean PN usage) was:
In 2016, 756 bags/month
In 2020, 603 bags/month
In 2024, 568 bags/month (25% reduction in PN provision from 2016)

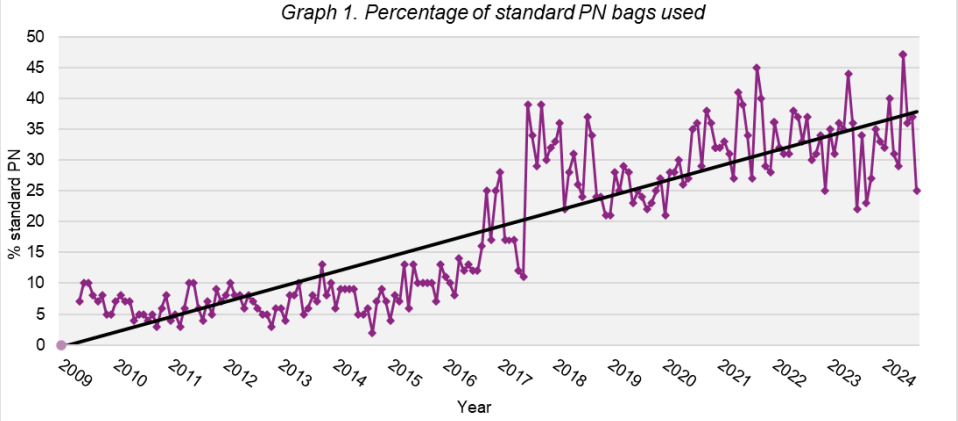
Standard PN usage – see Graph 1
Of PN provided, percentage of standard PN was:
In 2016, 14%
In 2020, 31%
In 2024, 35% (20% increase in standard PN usage since 2016)

PN wastage
In 2016, 3% of PN was wasted
In 2020, 4% of PN was wasted
In 2024, 5% of PN was wasted

NSIFT has had a continued positive impact on PN usage in the hospital. The team's interventions have resulted in lower PN usage and, with higher patient numbers, this suggests that patients have shorter episodes on PN. Individualised patient care plans offer better patient outcomes, reduce length of stay in hospital and provide significant cost savings. NSIFT continue to look for quality improvement opportunities to optimise PN, including timely weaning off PN and using standard PN more proactively. Using clinical expertise, the team have increased the portfolio of standard bag available. Interventions by the team have results in a 20% increase in standard PN usage from 2016 to 2024, which has resulted in at least £200,000 cost saving.

PN wastage is multifactorial, and there is still work to be done to achieve a sustained reduction in PN wastage. NSIFT are considering tools that can be implemented to support the appropriate prescribing of PN, including education and training to medica/surgery teams, tailoring support provided to teams and sharing the data collected to highlight the cost burden of wasted PN.

Graph 1. Percentage of standard PN bags used



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OC19

Small bowel enterostomy in the preterm infant: nutritional challenges and outcomes

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We aim to outline the experience of a Level 3 neonatal intensive care unit (NICU) in managing premature patients following small bowel enterostomy formation; to characterise the patient cohort, their respective outcomes and complications. Neonates <37 weeks gestation born between January 2021 and June 2024 undergoing small bowel enterostomy formation were included in the study. Data was collected retrospectively from electronic patient records and Microsoft Excel was used for statistical analysis.

28 eligible patients were identified; Table 1 summarises the cohort demographics. Median corrected gestational age (cGA) at stoma formation was 27.5 weeks (IQR 4.54 [25.97-30.5]) and median weight at stoma formation was 841 grams (IQR 398.25[650-1048.25]). Indication for laparotomy was predominantly documented as pneumoperitoneum (15/28; 54%); stoma site was predominantly ileal (21/28; 75%). 6/28(21.4%) patients died on NICU prior to stoma closure (median GA at birth 24.22 weeks (IQR 1.54 [23.61-25.15]); median birth weight 614 grams (IQR 110.5 [552-662.5]); 3/6 (50%) ileostomy, 3/6(50%) jejunostomy). 13/22(59%) surviving patients still had stomas at point of discharge and 2/22(9%) patients died following discharge whilst admitted to the paediatric critical care unit (PCCU) prior to stoma closure.

Only 4/22(18%) patients were documented to have achieved enteral autonomy (discontinuation of parenteral nutrition (PN) and effective growth (trajectory within 1 centile line of birth weight) on enteral feeding regime) prior to stoma closure or NICU discharge. Median GA at birth for these patients was 26.43 weeks (IQR 1.18 [25.64-26.75]) while cGA at stoma formation was 27.86 weeks (IQR 1.325 [27.68-29.0]), and median weight at stoma formation was 1015g (IQR 67.5 [972.5-1040]). All had an ileostomy and 3/4(75%) were discharged from NICU prior to stoma closure. A further 4/22(18%) patients achieved cessation of PN, however growth on enteral regime was not adequate.

Patients with small bowel enterostomy represent a significant workload within our NICU; experiencing a protracted inpatient stay, a challenging transition to enteral autonomy, and significant morbidity and mortality. These data support the importance of ongoing research into understanding the outcomes of neonatal patients post intestinal stoma formation and also may aid the prognostication of their likely nutritional course; allowing for effective family counselling and informing the transition to paediatric gastroenterology services.

Median Gestational Age at Birth (weeks)		25.79 (IQR 3.54 [24.14-27.68])
Median Birth Weight (grams)		656 (IQR 373 [589.5-962.5])
Location of Birth	Inborn	16(57%)
	Elsewhere	12(43%)
Corrected Gestational Age at Stoma Formation (weeks)		27.5 (IQR 4.54 [25.97-30.5])
Weight at Stoma Formation (grams)		841 (IQR 398.25 [650-1048.25])
Indication for Laparotomy	Pneumoperitoneum	15(54%)
	Suspected NEC	9(32%)
	Suspected intestinal atresia	2(7%)
	Small bowel obstruction	1(4%)
	Abdominal mass	1(4%)
Location of Stoma	Ileal	21(75%)
	Jejunal	7(25%)
Median Unit Days		104 (IQR 78.25 [42.25-120.5])
Median Unit Days Post Stoma		83 (IQR 94.25 [24.75-119])
Median Stoma Days on Unit		80 (IQR 75.5 [21-96.5])
NICU Deaths		6(21%)
Discharge Destination	PCCU	9(41%)
	Other Hospital/Ward	10(46%)
	Home	3(14%)
Discharged with Stoma	Yes	13(60%)
	No	9(41%)

Table 1 – Cohort Demographics

OC20

Dietetic service provision in paediatric intestinal failure: a national survey

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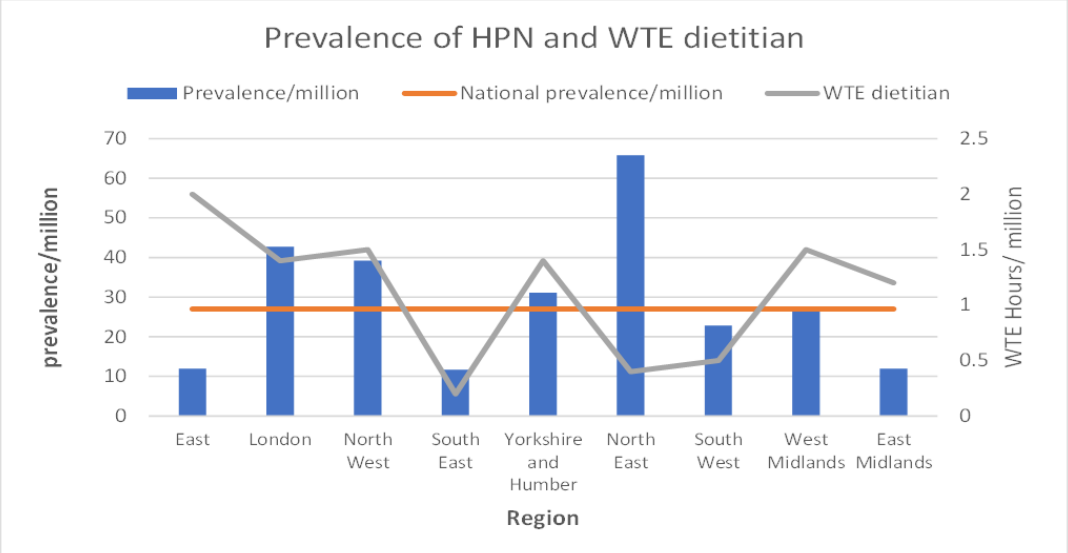
Management of parental nutrition (PN) is complex and multifactorial. The use of multi-disciplinary teams to manage intestinal failure (IF) patients is essential to ensure safe use of PN, promote enteral autonomy and improve outcomes (1-4). Gold standards for key members of paediatric IF team are published, yet little guidance exists for the whole time equivalent (WTE) that should be allocated for each member. To date there is no guidance for dietitians working in paediatric IF. The increasing prevalence rates and complexity of paediatric IF patients requiring long term PN (2-5) has wider implications for organisations and resources involved in the management of these patients. To help link resources and outcomes, job planning is essential to provide a means to understand workforce capacity and patient needs (6, 7).

This study aims to identify current service provision for paediatric dietitians in IF across England. It aims to explore if this provision is adequate to allow dietitians to meet all aspects of their job plans, set out by British Dietetic Association.

Two online surveys were sent to organisations providing paediatric IF service in England. Survey one focused on current dietetic service provision. Survey two concentrated on IF dietitian's workload demands, job plans and workload safety. To compare results to population, data was used from office National Statistics (ONS) mid 2022 dataset.

14/17 centres responded to survey one (82% response rate). The total number of paediatric patients receiving Home PN (HPN) in England was 326, with a prevalence rate of 27/million children, regionally prevalence rates ranged from 11.7/million to 65.8/million children. 50% centre respondents were able to specify exact WTE funding for paediatric dietetic IF services, the remaining centres provision came from gastroenterology funding, of which 40% were unable to specify how much time was specifically allocated towards IF. The total WTE dietitian provision nationally for IF services is 1.1/million children. Regionally, the WTE per head of paediatric population ranged from 0.2/million children to 2/million children.



Survey 2 had 10 respondents. 40% respondents had job plans. 80% felt they didn't have enough time to perform their job fully. 50% of participants, reported caseloads to be unsafe. 100% of participants work above their contracted hours with 60% working 1-5 hours extra, 30% working 6-10 hours and 10% working 11-15 hours over.

This study indicates that current paediatric IF dietitian service provision across NHS trusts in England is not sufficient. There is an apparent regional inequality of dietetic service provision, especially when compared to the prevalence of HPN within each area. All paediatric IF dietitians are working additional hours and the majority report unsafe workloads due to not having enough time to complete their job. With increasing IF patient complexity and numbers, further national guidance and research is required to develop standards.

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OC21

Splenic artery aneurysm in a patient with liver cirrhosis

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A 16 year old girl, with known advanced cirrhosis, portal hypertension and polysplenia, secondary to Biliary Atresia, post Kasai, presented with sudden onset left upper quadrant pain of one week duration. The pain was aggravated by lying flat, eating, coughing, mobilising and on deep inspiration. She required oral morphine for her pain. She reported her urine was more dark and her sclera were more jaundiced than normal. She was pyrexial 390C, tender in left upper quadrant with otherwise stable splenomegaly. There was no haemodynamic disturbance. Blood Investigations showed CRP of 79 (< 5); serum bilirubin 97 umol/L , ALT 48 (10-40); GGT 56 (< 38); ALP 168 (54-130). She had thrombocytopaenia 106, marginally improved to her usual level of 76 (secondary to hypersplenism). Prothrombin time was increased 19.1 (10.0-14.1) and haemaglobin was stable at 132 g/l. Lipase and chest x ray were normal.

In view history of previous Kasai, altered liver tests and pyrexia, she was commenced on IV Tazocin for possible cholangitis. An abdominal USS was requested to look for possible splenic infarct, which was not identified, although splenic varices were seen and portal vein was patent. After further discussion, a CT Angiogram mesenteric artery was requested, which showed multiple splenic artery aneurysms (SAA) ranging from 7mm to 27mm and presence of fluid around the region, concerning for a recent aneurysmal bleed.

She remained haemodynamically stable. Local Interventional radiology (IR), suggested embolisation of anuerysm(s) with consideration of splenectomy. She was well known to paediatric Liver centre and transferred for further assessment and management with a concern of potential acute liver decompensation if further aneurysmal bleed or secondary to intervention. She had a further bleed 10 days later and underwent repeat CT scan, which showed formation of a pseudoaneurysm and was embolised by IR, along with a few others. Following emobilisation she complained of left upper quadrant pain, likely due to splenic infarct but did not develop pancreatitis. There was a concern liver function was further compromised.

Reviewing the literature, SAA occurs more commonly in cirrhotic patients and are at a heightened risk of rupture peri liver transplant with potentially fatal consequences up to 57% mortality reported in one paper (1). However, there is no consensus regarding optimal management of asymptomatic SAA pre transplantation. A systematic review by Du Phan et al (2) reported on 159 patients with SAA, of whom 121 had asymptomatic SAA, diagnosed pre liver transplant and subsequently underwent liver transplantation. 37/121 patients diagnosed pre transplant had treatment instigated (28 surgically and 8 radiologically). Post-transplant rupture was noted in 2/28 patients treated surgically with no fatality. No rupture was observed in the radiologically treated group, although 1 patient died of splenic abscess and sepsis after embolization. Post-transplant rupture was recorded 4/86 untreated patients, (2/4 resulted in fatality). They concluded treatment should be considered regardless of aneurysmal size because of the risk of rupture post transplantation.

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OC22

Long-term maintenance of response and improved liver heath with maralixibat in patients with Progressive Familial Intrahepatic Cholestasis (PFIC): 2-year data from the MARCH-ON study

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Progressive familial intrahepatic cholestasis (PFIC) is a group of genetic disorders resulting in disrupted bile composition, cholestasis, and pruritus. Maralixibat, an inhibitor of the ileal bile acid transporter, is approved in the EU for the treatment of Progressive Familial Intrahepatic Cholestasis (PFIC) in individuals ≥3 months of age. In the 26-week placebo-controlled MARCH Phase 3 study, MRX at 570 µg/kg BID demonstrated significant improvements in pruritus, serum bile acids (sBA), total bilirubin (TB) and growth in patients across the broadest range of PFIC types studied to date. We report on long-term maintenance of effect of up to 2 years of treatment with MRX in MARCH-ON, an open-label, long-term extension study of MARCH.

Long-term maintenance of response was assessed for patients who were originally randomized to receive MRX in MARCH and continued with treatment in MARCH-ON (MRX-MRX group: n=47), and for patients who received placebo (PBO) in the MARCH study and switched to open-label MRX in MARCH-ON (PBO-MRX group: n=41). Assessments included: pruritus, sBA, TB, growth z-scores, and incidence of treatment-emergent adverse events (TEAEs). Baseline (BL) was defined as the start of MRX for each group.

For the MRX-MRX group, the median (min, max) time on MRX was 638 days (10, 1135). 13 of 47 patients reached Week 104 at time of analysis. Significant improvements observed in the first 26 weeks of the MARCH study were sustained through Week 104 in MARCH-ON for pruritus (-2.03, $p<0.0001$), sBA (-166 µmol/L, $p=0.003$), TB (-27.7 µmol/L, $p=0.02$), and growth (height z-score: +0.40, $p=0.046$; weight z-score: +0.52, $p=0.01$). In the PBO-MRX group, the median time on MRX was 456 days (22, 720). 18 of 41 patients reached Week 52 of MRX treatment at time of analysis. Significant improvements through Week 52 for pruritus (-1.1, $p=0.0001$), sBA (-71 µmol/L, $p=0.03$), and growth (height z-score: +0.37, $p=0.01$; weight z-score: +0.32, $p=0.03$) were in line with observations from the initial MARCH MRX group. Additionally, numeric reductions in TB (-6.4 µmol/L; $p=0.7$) were observed. No new safety signals were identified. The most common TEAEs were GI-related with diarrhea (50%) being mostly mild and transient.

Significant and sustained improvements in pruritus, sBA, TB, and growth are observed with up to 2 years of MRX treatment across the broadest range of genetic PFIC types studied to date. These data suggest overall improved liver health with MRX treatment which can be maintained long-term.

OC23

Case Report: Acute Liver Failure following Exercise-related heat illness in a British Teenager

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Exercise related heat illness (EHI) in young persons is rare in temperate regions. We report a case of EHI in a teenager who presented with acute liver failure (ALF), acute kidney injury (AKI) and rhabdomyolysis following vigorous physical activity. We also highlight the challenges of interpreting ammonia results if liver transaminases are elevated.
A 15-year-old British male fell unconscious while on a 10km run on a warm humid day. The weather conditions at the time of the run were 23 °C, humidity of 55%, and 6mph wind speed. A passerby finding him pyrexial, dowsed him with water and moved him to a shaded area. Past medical history included well controlled asthma and a holiday to Egypt 2 weeks back.

Paramedics on arrival noted tympanic temperature of 39.1 °C, tachycardia and delirium. He received fluid boluses and was intubated for transfer but extubated shortly after arrival to the hospital. Initial bloods showed deranged liver function, kidney tests and high troponin and creatinine kinase (CK), (Figure 1). Full blood count was within normal limits. CT head was unremarkable and urine toxicology screen negative.

He remained well and on day 3, while awaiting cardiac investigations was noticed to be jaundiced. His repeat bloods showed ALF with an International normalized ratio of 3, which remained unchanged despite parental Vitamin K. With an AKI, increasing increased ammonia and CK (Figure 1), ALF protocol and N acetyl cysteine (NAC) infusion were initiated prior to transfer to the specialised liver centre.

On arrival to the liver centre 76 hours post the initial insult, he had normal vitals and sensorium. Blood tests for liver and Kidney injury were beginning to improve and a normal ammonia was recorded (Figure 1). Etiological work up for other causes of liver injury were negative (Infection, metabolic, autoimmune, genetic including mitochondrial diseases). He made a full recovery with supportive care and NAC infusion. Further genetic testing for malignant hyperthermia is negative.

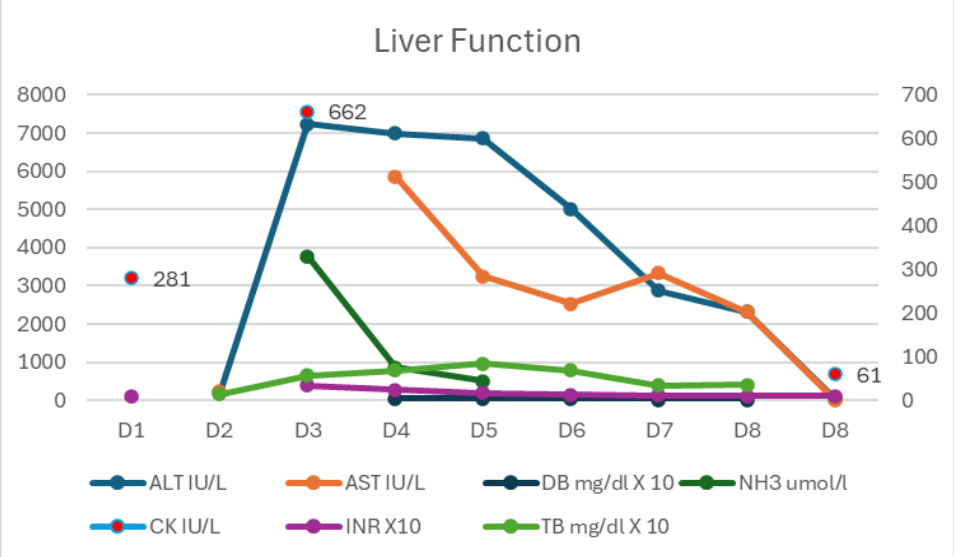
With the increasing risk of EHI due to climate change, this case highlights the importance of recognition, early cooling and need for blood test monitoring as severe organ injury may not be evident at onset[1]. Ammonia may be inaccurately recorded high as in this case, if tested by wet enzymatic assay in presence of elevated liver transaminases (as at the admitting hospital). This has therapeutic implications as hemofiltration may be unnecessarily instituted. Laboratory wet chemistry ammonia analysis should include adequate sample blanking correction for accurate results[2] (as at Liver unit) or be undertaken by dry chemistry methods.

World Health Organisation recommends NAC for heat stroke induced ALF and may help by replenishing glutathione stores[3]. Genetic testing for mutations that increase EHI susceptibility aids counselling[4]. Assessment in controlled environment by sport physiologist is recommended for advice on heat acclimatisation, establishing a prevention plan and safe limits of physical activity for the individual[5].

Figure 1: Liver biochemical tests during course of illness

Primary axis: ALT, AST

Secondary axis: Rest



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OC24

Role of sarcopenia in paediatric metabolic dysfunction-associated steatotic liver disease (MASLD).

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Studies in adults have demonstrated that the presence of sarcopenia in patients with MASLD is associated with a higher likelihood of steatohepatitis and advanced liver fibrosis independent of other confounding factors like age, sex, BMI and insulin resistance.

Children diagnosed with MASLD undergoing liver biopsy were recruited to have body composition assessed by DXA scan and bioelectrical impedance (BIA). The participants also had HOMA-IR calculated as well as standard anthropometry and blood tests, ELF score, physical activity questionnaire score. Liver biopsies were assigned NAS score and fibrosis stage.

Twenty children were recruited. Age 10-16 years (16M:4F). Median z-scores were for weight 2.69 (range 0.89 to 3.46), for height 0.86 (range -0.17 to 2.87) and for BMI 2.31 (range 1.04 to 3.56). Eighteen children of these had whole body DXA scans, all had BIA. None of the children were sarcopenic as per definition of fat-free mass index (FFMI) z-score less than -2. All indices correlated with each other strongly (weight, BMI, total FMI, total FFMI, appendicular FMI and FFMI z-scores). Mean z-scores for FFMI were significantly different to the corresponding FMI z-scores of the same child.

NAS scores of liver biopsies: 2 scored 1, 4 scored 2, 6 scored 3, 7 scored 4 and 1 scored 5. Biopsies with a score equal or less than 3 were no MASH, over 3 with MASH. Fibrosis was staged as 0, 1a, 1b, 1c, 2, 3, and 4. No children had fibrosis stage 4, 5 had stage 3, 5 had stage 2, 5 were stage 1 and 3 were stage 0. Those with stage 0-1 we grouped as mild fibrosis, stage 2 and 3 were grouped as advanced fibrosis. The children classified as MASH had a higher cholesterol/ HDL ratio, but no significant anthropometric differences. The children with the more advanced fibrosis also had a higher cholesterol/HDL ratio but no anthropometric differences. In the advanced liver fibrosis group there was a tendency for more children to have a FFMI z-score in the lower quartile (75% vs 25%) on liver biopsy, but the numbers were small for more definite conclusions.

%Lean mass (%LM) derived from the BIA correlated strongly with the FM indices from the DXA, implying that as fat mass increased the %LM was less, without necessarily an actual reduction in LM.

HOMA-IR had a significant negative correlation with total body FFMI z-score -0.609 and other expressions of FFMI like (FFMI/FMI z-score or FFMI/BMI z-score).

In our cohort of patients with biopsy proven MASLD, none of the patients were sarcopenic per se. HOMA-IR had a significant negative correlation with FFMI z-score. There was a tendency for the group with the more advanced liver fibrosis to have lean mass in the lower centiles.

OC25

Rising incidence of paediatric autoimmune liver disease – ? an immune mediated post-COVID-19 phenomenon

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Higher incidences of type 1 diabetes and coeliac disease are increasingly reported compared to pre-COVID-19 pandemic levels[1,2]. Moreover, higher rates of severe diabetic ketoacidosis are seen in children with newly diagnosed diabetes[1,3].
The incidence and disease severity of paediatric autoimmune liver disease (AILD) between pre and post-COVID-19 pandemic levels has not been assessed. Retrospective data from a cohort of 202 children with newly diagnosed AILD over a 10 year period between 2015 to 2024 was reviewed from a UK quaternary liver specialist centre.

Findings show a higher incidence of autoimmune hepatitis type 1 (AIH-1), Primary sclerosing cholangitis (PSC) and Overlap/Autoimmune sclerosing cholangitis (ASC) in the post-COVID-19 time period (2020-2024) compared to pre-COVID (2015-2019), with the greatest increase observed in the AIH-1 cohort (p<0.0001) (Figure 1). A higher age-range for all three subgroups was seen but most notably in AIH-1 (p=0.0093). In contrast, minimal variation for incidence and age-range was observed in AIH-2 patients. The INR was higher in AIH-2 patients compared to pre-pandemic levels (p=0.0198).

A greater proportion of **patients** presenting with decompensated liver disease was observed in all subgroups post-COVID-19 but was **most marked** in Overlap/ASC children (p=0.0313). Next the development of IBD (**≤1year of AILD diagnosis) was assessed; higher rates of new-IBD was observed in PSC patients post-COVID-19** (p<0.0001), specifically for Crohn's disease (p=**0.**0030) and indeterminate colitis (***p***<0.0001). Patient demographics were interrogated to identify possible associated factors; a significant rise in the incidence of African patients presenting with AIH-2 (p=0.0**011**) and Overlap/ASC (p=**0.**0043) was observed post-COVID-19. The rate of concomitant viral infections including COVID-19 was low in all subgroups; no patients experienced severe COVID-19 or PIMS-TS.

In conclusion, our study is the first to describe a higher incidence of children presenting with newly diagnosed AILD in the post vs. pre-COVID-19 pandemic era. Importantly, children presented older, had a higher rate of decompensated liver disease and patients with PSC were more likely to develop concomitant IBD.

The higher incidence and disease severity observed in our AILD cohort and that reported in the literature for other autoimmune conditions[1-3] maybe a post-COVID-19 pandemic phenomenon resulting in an abnormal immune-response in susceptible individuals following on from long-periods of social isolation and the lifting of COVID lock-down measures.

Figure 1.
Line graph displaying incidence of AIH-1 – the dotted line splits the graph into pre-COVID-19 (2015-2019) and post-COVID-19 (2020-2024) time periods. The incidence of
AIH-1 rises sharply from 2020 and peaks at 2021 prior to decreasing
to an incidence level which remains above pre-COVID-19 baseline levels.

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OC26

The relationship between serum bile acids and event-free survival following the use of maralixibat for Progressive Familial Intrahepatic Cholestasis: Data from MARCH/MARCH-ON

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Maralixibat, an inhibitor of the ileal bile acid transporter, is approved in the EU for treatment of Progressive Familial Intrahepatic Cholestasis (PFIC) in individuals ≥3 months. Prior analyses in ALGS have demonstrated improved event-free survival (EFS) following use of maralixibat and this improvement is associated with reductions in sBA. Data from MARCH/MARCH-ON, a clinical trial of maralixibat for individuals with PFIC, demonstrated reductions in sBA. In this analysis, we evaluated the impact of sBA reduction on EFS.

The design of MARCH/MARCH-ON have been previously described. Data were administratively censored in June, 2023. First events (i.e., liver transplant, decompensation, surgical biliary diversion [SBD], or death) were identified for different PFIC types. For individuals with nt-BSEP and FIC1, 2-year EFS was calculated and stratified by sBA response at Week 26 (averaged over last 12 weeks) using thresholds developed by the NAPPED Consortium (BSEP: >75% reduction or <102 µmol/L; FIC1: <65 µmol/L).

There were 5 events (nt-BSEP: 1 transplant, 1 decompensation; FIC1: 1 death, 1 SBD; and MDR3: 1 transplant) among 72 individuals with a median (Q1, Q3) follow-up of 94 (68, 110) weeks. The overall EFS was 92%. Among individuals with nt-BSEP, an sBA response was achieved in 12 of 27 (44%), and this group had no events yielding an EFS of 100%; an insufficient sBA response was observed in 15 (56%) and this group had 2 events yielding an EFS of 84%. The sBA reduction for the 2 patients with events were 19% and 26%. Among individuals with FIC1, an sBA response was achieved in 3 of 12 (25%) and this group had no events yielding an EFS of 100%; an insufficient sBA response was observed in 9 (75%) and this group had 2 events for an EFS of 78%. The sBA reduction for the 2 patients with events were 18% and 16%. When nt-BSEP and FIC1 were analyzed together, sBA responders had an EFS of 100% whereas sBA non-responders had an EFS of 81%. For the individual with MDR3 disease requiring a transplant, the sBA reduction was 44%.

Consistent with sBA response thresholds from the NAPPED Consortium that are associated with EFS, individuals in MARCH/MARCH-ON who reduced sBA levels below the threshold did not have a clinically meaningful event whereas some individuals who had lower reductions in sBA experienced events. These data support the importance of sBA reduction in PFIC and the potential of maralixibat to facilitate this biochemical change.

OC27

Rare but treatable cause of cholestasis- primary bile acid synthesis defect

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Bile acid synthetic defects are uncommon genetic disorders causing persistent cholestasis in infants. The onset and severity of the liver disease is extremely variable from mild, transient, and delayed in onset, while on the other end of the spectrum, presenting with severe neonatal or infantile cholestasis with progression to early cirrhosis and liver failure necessitating liver transplantation or leading to mortality.

Patient was incidentally detected to have jaundice and significantly raised transaminases (> 1000 IU/ml) at the age of 5 months. She was born to non-consanguineous Caucasian parents. At the time of presentation itself metabolic or genetic disorder was considered as a strong possibility as there was massive hepatomegaly, splenomegaly, normal gamma glutamyl peptidase level and diffusely bright liver in ultrasound imaging. Initial assessment for metabolic disorders including tyrosinemia was negative. The serum bile acids were normal. The alpha fetoprotein level was significantly raised and worryingly, showed a rising trend. This prompted further assessment with abdominal magnetic resonance imaging in which showed multiple bilateral T1 hypointense, T2 hyperintense lesions with restricted diffusion with no central enhancement involving both kidneys.

Liver biopsy showed features of chronic liver injury with early cirrhosis. Genetic testing identified heterozygous pathogenic variant (*HSD3B7 c.45-46del p. (Gly17fs)*) in the HSD3B7 gene. Urinary cholanoids by electrospray ionisation tandem mass spectrometry detected sulphated dihydroxy- and trihydroxy-cholenoic acids present as major peaks confirming diagnosis of 3-beta-hydroxy-delta5-C27-steroid dehydrogenase deficiency (3β-HSD), the most common type of bile acid synthetic defect. Patient was started on oral cholic acid therapy. At 2 years of follow-up, cholestasis, hepatomegaly, splenomegaly and renal lesions have resolved, child is growing well with normal developmental milestones.

Bile acid synthetic defects are caused by defective enzymes catalyzing key reactions in the formation of the primary bile acids (cholic acid & chenodeoxycholic acid), causing inadequate synthesis of primary bile acids which are critical for bile formation, absence of these leads to the accumulation of atypical and hepatotoxic bile acid intermediates. Absence of pruritus, normal serum gamma glutamyl transferase activity, and normal or low total serum bile acid concentration are diagnostic clues. Care is required when interpreting a serum bile acid level obtained when the patient is receiving ursodeoxycholic acid because elevated serum bile acids may not necessarily exclude a diagnosis of HSD3B7 deficiency in such patients. Renal lesions, most commonly renal cysts, but also renal stones, calcium deposition and renal enlargement are observed in 28% at the time of presentation. The diagnosis is confirmed by mass spectrometry analysis of urinary bile acids showing typical bile acid profiles and genetic analysis.

Lifelong oral therapy with bile (cholic) acid is safe, effective with complete normalisation of liver biochemistry, radiological abnormalities and renal lesions. Cholic acid therapy has been shown to reverse the extent of fibrosis. Regular monitoring of urinary bile acid profile is crucial to assess treatment efficacy and compliance

OC28

Incidental finding of High Rates of Learning and Behavioural Difficulties in children living with Hepatitis C

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Background

Chronic HCV infection is asymptomatic in CY&P with rare cases of advanced fibrosis & hepatic failure . The central nervous system impact of HCVin adults is well described, with both systemic immune activation and viral replication implicated.[1] There has been little focus on neurodevelopmental data for CY&P living with HCV, yet they face exposure to both maternal systemic inflammation in utero and to HCV & immune activation throughout the critical period of infant brain growth in the first 2 years of life.

Maternal inflammation from both infective & inflammatory triggers is associated with an increased risk of neurocognitive & psychiatric disorders in childhood[2]. Additionally, infants born to women living with HCV are more likely to experience in utero substance use exposure, with the potential for further neurocognitive sequalae associated with neonatal abstinence & fetal alcohol syndromes.[3]

Neurocognitive deficits in children appear to correlate with levels of plasma inflammatory cytokines, including comprehension and vocabulary.[4] The impact of DAAs on childhood neurocognitive functioning, requires elucidation.

Methods

Retrospective review of e-referrals to HCV pMDT ODN between 04/21 to 09/24
Data collected:
•Clinical & demographic data ; • Co-morbidities; •Neurodevelopmental & behavioural difficulties (after 04/23) •Issues affecting adherence; •DAA formulation; •Outcomes
Comparative data reported:
Group 1: No reported learning & behavioural difficulties (NLD); **Group 2:** Reported learning & behavioural difficulties (LD)

Results

141 HCV+ve C&YP referred to the pMDT ODN:
•Group 1 n= 112 (NLD). •Group 2 n-29 (LD)

Table 1 Comparative data

	Group 1	Group 2
Genotype • 1 • 2 • 3 • 4	 54 5 49 4	 13 1 14 1
Median (range) age years	8.6 (3 - 17.8)	8.1 (3.2 - 13.3)
Gender	61M	13M
Ethnicity • Caucasian • Asian • Unknown • African • Others	 64 21 14 4 9	 26 2 -- -- 1
Transmission • Vertical • Horizontal • Unknown	 107 2 3	 28 1 --
Formulation prescribed • Tablets • Granules	 77 35	 17 12

OC28 - continued

Table2: Treatment outcome Group 1 v Group 2

	Group 1	Group 2
Treated	99	23
Treatment milestones* • End of Treatment • SVR12 • 12mSVR	 90 82/90 61/82	 21 16/21 10/16
Treatment not started • Recently approved • Awaiting feedback • Family no longer in UK • Child refusing treatment	 4 6 1 2	 2 0 0 4
Blood monitoring • None • Child refusing** • Dificulties obtaining bloods** • DNA appointments*** • Lost to follow up***	 96 1 0 1 1	 17 1 1 4 --

*Outcome where specific end point has been reached

**Clinicians working with support teams

***Working with health agencies to locate families

Table 3: Summary of findings in Group 2

Findings	
Behavioural/Learning difficulties	4/4
Foetal Alcohol syndrome/neonatal drug withdrawal	7/6
Attention Hyperactivity Deficiency/Autistic Spectrum Disorder	9/12
Global developmental delay	4

Summary:
Learning & behavioural difficulties were reported in 21% compared to estimated rates of 2.5-4% in the general UK school aged population. There was no significant difference in demographics or treatment response; children with LD had more difficulty attending clinic and taking the medication.

Conclusion
The potential role of maternal-infant HCV infection on neurocognitive outcomes requires elucidation for optimal timing of HCV therapy, currently not licensed until age 3+ years & to determine the risk benefit of maternal HCV clearance in pregnancy.

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OC29

Clinical benefits of maralixibat for patients with Alagille syndrome are durable through 7 years of treatment: data from the MERGE study

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Maralixibat (MRX), an ileal bile acid transport inhibitor (IBATi), is approved for the treatment of cholestatic pruritus in patients with Alagille Syndrome (ALGS) ≥2 months of age in Europe. Improvements in pruritus, serum bile acids (sBA), and height have been demonstrated from prior clinical trials including ICONIC, which followed participants up to 4 years, as well as IMAGO/IMAGINE and ITCH/IMAGINE-II, which reported outcomes to approximately 1.5 years. Participants from ICONIC, IMAGINE, and IMAGINE-II trials were invited to enroll in MERGE for additional long-term follow-up (LTFU); prior long-term survival outcomes (e.g., liver transplant, death) for this group have been previously reported. Here we report on efficacy in participants with additional LTFU from MERGE, including some participants that have received treatment for 7 years.

All participants from ICONIC, IMAGINE and IMAGINE-II were included in the analysis. Impact of MRX was assessed for pruritus [ItchRO(Obs) 0-4 scale, with a ≥1-point reduction considered clinically meaningful], sBA, height and weight z-scores, ALT, total bilirubin (TB) and direct bilirubin (DB). Change from Baseline (CFB) was determined by comparing median (Q1, Q3) values from enrolment in the initial trial (i.e., ICONIC, IMAGO, or ITCH) to data from the visit in MERGE that best aligned with an annual visit.

Data were analyzed for 86 participants at Baseline, with follow-up to 1 year for 76 participants, 4 years for 42 participants, and 7 years for 23 participants. Of the 86 participants, 84 had a genetic diagnosis of ALGS via the *JAG1* mutation, 2 participants had a genetic diagnosis of ALGS via the *NOTCH2* mutation, and 1 participant had an unidentified mutation. Baseline mean (SD) ItchRO(Obs) was 2.65 (0.75) and clinically meaningful reductions over time with CFB of -1.57 (-0.83, -2.14), -2.00 (-1.43, -2.56), and -2.14 (-1.43, -3.00) at 1 year, 4 years and 7 years, respectively. Likewise, Baseline sBA was 254 (207) µmol/L and decreased with CFB of -57 (8, -150) µmol/L, -62 (-32, -152) µmol/L, and -105 (-41, -266) µmol/L at 1 year, 4 years and 7 years. Improvement was observed in height, with Baseline z-score of -1.7 (1.27) and CFB of 0.1 (-0.1, 0.3), 0.3 (0.0, 1.0), and 0.7 (0.0, 1.2) at 1 year, 4 years and 7 years while weight z-scores were largely unchanged. Reductions in TB and DB were observed after treatment with maralixibat. No clinically meaningful changes in ALT or AST were observed with maralixibat treatment. There were no new safety signals.

In this unmatched cohort, the benefit of MRX in ALGS patients, including both improvements in clinical outcomes and sBA, persist through 7 years of treatment. No new safety concerns were identified in the long-term.

OC30

Single centre tertiary paediatric gastroenterology unit experience of Ustekinumab

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Ustekinumab is an anti IL12 and IL 23 monoclonal antibody available to treat Crohn's and Ulcerative colitis. We wanted to review it's use in our patients. A list of patients was obtained from "Infoflex" database and electronic patient notes were reviewed for IBD type; age at a diagnosis; age of starting Ustekinumab; disease distribution; previous biologics and outcome of our patients who had been treated with Ustekinumab. 27 patients were treated with Ustekinumab, of whom 2 received Ustekinumab on 2 separate episodes. 4 patients were excluded as they had transferred to adult services with no follow up data. 23 patients; (10 females) were evaluated. All patients received an induction dose intravenously followed by subcutaeous injection as per standard protocol.21 patients were diagnosed with Crohn's disease (13 ileocolonic disease) and 2 had IBDU. The median age at diagnosis was 13.8y (range 3.5y- 15.1y). The median age of starting Ustekinumab was 11.7y (range 11.3y – 16y). All patients had undergone conventional treatment with steroids and or exclusive enteral nutrition; azathioprine and between 1 and 3 biologics, including adalimumab, infliximab and vedolizumab, prior to starting Ustekinumab. 12/21 patients stopped treatment because of poor response. 4/12 patients underwent surgery for failed medical treatment; 2 subtotal colectomy with ilesotomy and 2 right hemicolectomy. Ustekinumab drug levels were not freely available. 5/12 patients stopped Ustekinumab because of inadequate response, despite adequate drug levels of > 1.1mg/L. Only 2/23 patients reported side effects of Ustekinumab.Ustekinumab was a safe and well tolerated drug, although more than half the patients stopped treatment due to inadequate clinical response. Proactive drug therapeutic monitoring may help improve our understanding of appropriate dosing of Ustekinumab and levels required to treat paediatric patients optimally compared with adult patients.

OC31

Response to exclusive enteral nutrition in children with Inflammatory Bowel Disease specifically any differences with ethnicity, demographics rather than just phenotype

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Ultra-processed foods (UPFs) are widely consumed due to their affordability and convenience, yet they are associated with adverse health outcomes, including gastrointestinal diseases.(1) The incidence of Crohn's Disease (CD) has been increasing in regions with rising UPF intake.(2) This study aims to evaluate dietary factors, specifically UPF consumption, in relation to remission duration and demographic factors in paediatric CD patients following successful exclusive enteral nutrition (EEN) induction. The primary objective is to determine the relationship between UPF intake, captured via a one-time food frequency questionnaire, and remission duration (days) in this cohort. Secondary objectives include analysing UPF consumption across ethnicity and socioeconomic backgrounds.

We conducted an observational cohort study in CD patients (≤18 years) diagnosed between April 2023 and April 2024 and successfully treated with EEN at a specialised centre. Data were collected via a questionnaire and the patients' electronic records, and statistical analysis was performed in IBM SPSS statistics version 29.

11 participants (six female) were included in the cohort; median (IQR) age was 13 (7-18), 54.5% had ileal disease, 100% had a non-stricturing non-penetrating phenotype, 45% had upper GI disease,18.2% had perianal disease, 45.4% were Asian/Asian British, and 63.6% were from low socioeconomic backgrounds. Six patients relapsed post-EEN, and mean remission length was 62.7% lower in the relapse group. Table 1 summarises the cohort's dietary patterns. Mean UPF consumption was higher in the relapse group (42.30, SD 18.96) compared to the remission group (38.64, SD 13.95), but no statistically significant association was found between UPF intake and remission duration (Cox regression, fully adjusted model: HR 0.93, 95% CI 0.772–1.116, p=0.429). Higher mean UPF consumption was associated with higher socioeconomic status (high; 46.59, SD 7.23, middle; 39.79, SD 28.22, low; 39.18, SD 16.71) and Black/Black British ethnicity (Black/Black British 47.01, SD 15.28, Asian/Asian British 36.68, SD 20.59, Other 40.84, SD 11.19), however this was not statistically proven (Kendall's Tau correlation coefficient for socioeconomic status 0.191, p=0.463, Kruskal-Wallis, mean ranks; Black/Black British 7.33, Other 6.55, Asian/Asian British 5.00, p=0.970).

In conclusion, higher UPF intake was not statistically associated with remission length in children with mild-to-moderate CD post-EEN induction. UPF intake didn't vary substantially across socioeconomic or ethnic groups. Larger longitudinal studies are needed to clarify the role of UPFs in CD progression before introducing dietary guidelines in clinical practice.

Table 1 Cohort's Dietary Patterns and UPF Consumption

Variables		**Total (n=11)**
BMI (kg/m^2)	Median (Range)	20.2 (14.3-24.8)
BMI-for-age (percentile)	Median (Range)	75.0 (12.0-98.1)
BMI-for-age percentile category (n) *Underweight* *Healthy Weight* *Overweight* *Obesity*		 - 9 (81.8%) 1 (9.1%) 1 (9.1%)
Total energy consumption (kcal/day)	Median (Range)	1878.2 (1198.5-2244.6)
UPF consumption (kcal/day)	Median (Range)	694.2 (297.9-1411.5)
UPF consumption (%)	Median (Range)	41.1 (17.2-68.5)
Fast food/take away consumption prior to CD diagnosis (servings/day)	Mean (SD)	0.35 (0.36)
Fast food/take away consumption post CD diagnosis (servings/day)	Mean (SD)	0.18 (0.27)
Data are presented as n (%) or median (range) / mean (SD). BMI measured after EEN completion.		

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OC32

Reviewing the psychological needs of patients with Inflammatory Bowel Disease: A retrospective study within a Paediatric Gastroenterology Psychology Service

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Psychological support for children and young people (CYP) with Inflammatory Bowel Disease (IBD) is integral to supporting optimal wellbeing (1). The biopsychosocial nature of IBD means that psychological support is central in helping patients and families understand the condition, cope with the treatments and achieve optimal psychological functioning. However, psychology is often a limited resource within IBD teams (2).
CYP with IBD comprise a significant proportion of referrals to the Specialist Gastroenterology Psychology Service. This study aimed to provide a retrospective analysis of referrals to identify key reasons for referral, consider patterns of referral and better understand the need for psychology support.

A review of psychology referrals made for CYP with IBD between October 2023 and October 2024 was undertaken. This study analysed demographic information, reasons for referral and general treatment details including waiting times and sessions attended.

66 CYP with IBD were referred to the Paediatric Gastroenterology Psychology Service between October 2023 and October 2024, with most (76%) being outpatient referrals. CYP with IBD represented 38% of referrals during this period. Interestingly, there were an additional 16% of CYP referred who did not engage with the service after referral, of which 100% were outpatient referrals. For CYP who did engage, there was a wait to be seen that spanned 36 and 96 days, with an average wait of 59 days. The wait for inpatient support was shorter, between 3-18 days. Age at referral ranged between 3 and 17 years, with an average referral age of 11. The number of treatment sessions ranged between 2 and 16 sessions, with an average of 4 sessions.

The most common reasons for outpatient referral were for support with adjustment to diagnosis, high anxiety connected to IBD and treatment adherence. In terms of inpatient referrals, the most common referrals were for support with adjustment to diagnosis, procedural distress and surgical preparation, high anxiety and low mood. The data suggests inpatient support required longer and more frequent sessions to best support families.

The findings suggest that overall, psychology support is vital in the management of CYP with IBD in this service. Clinical psychology service provision should be developed with the needs of the CYP in mind, considering more efficient referral processes and ways of increasing effectiveness with limited resources.

The differing needs of the groups should be considered, and the service streams developed accordingly. For outpatients, there is a need to investigate barriers to engagement with the psychology service. In addition, groups aimed at supporting adjustment and adherence could provide efficient and effective psychological support.

In terms of inpatient support, the need for a fast response from the point of referral coupled with the need for more intensive support suggests that practical strategies, such as having a nominated clinician with protected time, could work well in providing optimal support.

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OC33

Comparison of Anti-TNF treatment in paediatric patients with ulcerative colitis: Adalimumab vs Infliximab in a five-year retrospective audit from a regional cohort

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Current paediatric guidelines recommend Infliximab as the first-line anti-TNF therapy for patients with moderate to severe Ulcerative Colitis (UC)1 and Adalimumab as second-line if there is loss of response2,3. Some adult studies, however, have shown similar efficacy when comparing both in maintenance treatment4,5. Due to bed availability, treatment duration, and to reduce disruption to schooling/family life with subcutaneous injections instead of intravenous infusions, our specialised centre's protocol is to initiate Adalimumab as first-line for most UC patients when clinically appropriate.

This audit aims to assess whether it makes a difference to the treatment outcome for patients with UC which anti-TNF medication is started; and on which treatment the patient remained until adult transition (or up until data was collected); the rationale for starting Infliximab as opposed to Adalimumab; why the medication switch was initiated, and review both the Adalimumab induction and maintenance doses.

We conducted a retrospective audit in our specialised centre for newly diagnosed UC patients from 1/1/2019 to 31/12/2023. We analysed the data of 186 children (107 males:79 females; aged 3 months to 17 years, Mean 11.64y, Median 12y), of which 87 were started on Anti-TNF treatment (64 Adalimumab vs 23 Infliximab) (Figure 1).

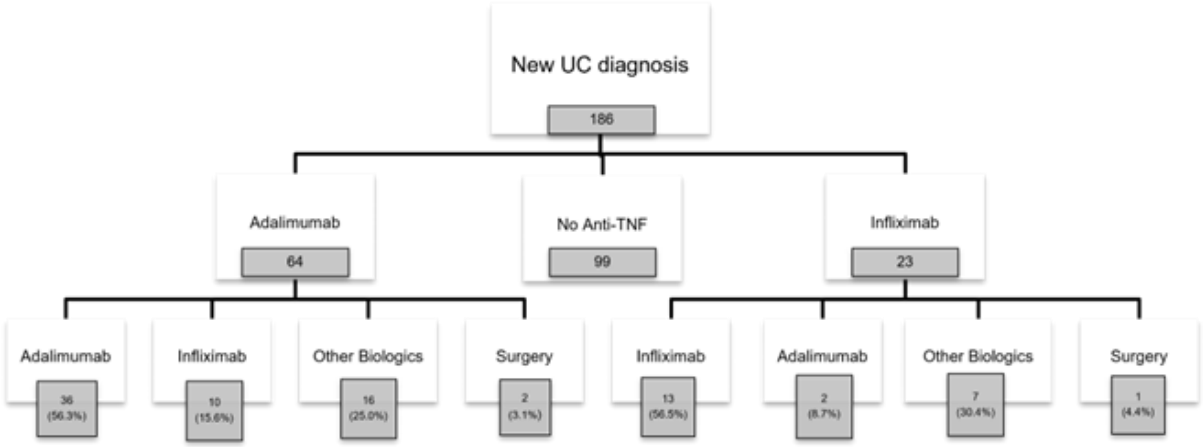

Figure 1: Representation of initial treatment groups and subsequent outcomes

The data showed the two Anti-TNF groups lead to similar percentages of patients remaining on the same first-line treatment (Adalimumab 56.3% vs Infliximab 56.5%) with statistically not significant difference in comparison of proportion (difference 0.2%, 95% CI -22.6% to 21.8%, p=0.9868) as well as relative risk (RR=1.00, 95% CI 0.58 to 1.73, p=0.9820) and odds ratio (OR=1.01, 95% CI 0.38 to 2.64, p=0.9820).

Moreover the difference in comparison of proportion was not statistically significant when the patients switched to the other Anti-TNF (p=0.4129), to other biologics (p= 0.6165) or had surgery (p=0.7706).

The analysis also demonstrated the main reasons to start on Infliximab instead of Adalimumab (N=23) were symptomatic patients with high faecal calprotectin (FC) (65.2%) or acutely admitted patients (34.8%).

The data demonstrated that patients were mainly switched from Adalimumab to other treatments (N=28) due to non-response (35.7%), loss of response (25.0%) or relapse (21.4%).

Furthermore, 85.7% of patients who started on Adalimumab were given the loading doses as per guidelines1, and the ones who remained on this Anti-TNF had a 55.6% vs 44.4% split between weekly and 2-weekly injections.

In conclusion, using Adalimumab as first-line Anti-TNF treatment for paediatric UC patients appears to yield similar outcomes to Infliximab. Given these findings, weighing cost-effectiveness and the benefits to the patients our specialised centre has continued with its current protocol, and other specialised centres may consider similar practice. However further studies are needed to determine whether these outcomes are driven by intrinsic biological factors or logistical treatment variables.

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OC34

Single-centre experience in using Upadacitinib in moderate to severe paediatric inflammatory bowel disease in a tertiary paediatric centre.

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Upadacitinib (UPA) was recently approved for use in Ulcerative colitis and Crohn's disease in adults, Despite the promising data in the adult field, still limited data available for use in paediatrics. (1) Upadacitinib is a Janus kinase inhibitor licensed for adult IBD and possesses numerous benefits of being a small molecule, oral administration, and no immunogenicity. (2) Upadacitinib targets JAK1, thus limiting immune cell adhesion and release of cytokines. (3)

We identified patients who were <= 18 with refractory IBD who were started on Upadacitinib (UPA). The data collected from the electronic records included the type of disease and its extent, age at diagnosis and age at starting UPA, duration of the disease, what biologics failed before, what the outcome of treatment based on symptoms, blood test (including inflammatory markers (CRP, Albumin, faecal calprotectin).

We had six patients with refractory IBD who received UPA,50% were male. The age at the start of UPA ranged from 14.5y-18ys (mean age 13.3 years), with a disease duration ranging between 4.5ys -10ys (mean 6.3 years).

All the patients had Crohn's disease; four had the non-structuring disease (B1) (three of those had ileocolonic and one had colonic disease), and the remaining two had a perianal fistulating disease.

All the patients were biologic-exposed, (60%(n=4) were exposed to three previous biologics), and the 40%(n=2) had two previous biologics. Before initiating UPA 83%(n=5) received infliximab, 50%(n=3) had adalimumab,50% Vedolimumab,66%(n=4) had Ustekinumab,66%(n=4) had Azathioprine. All patients received UPA as monotherapy.

UPA induction was administered for 12 weeks at a dose of 45mg once a day, then reduced to 15mg as maintenance. Only one patient was kept on 30 mg due to chronic renal disease. Duration on UPA use ranged from 6 months to 16m (Median 8m). Three patients were transitioned to adult services. One patient was excluded due to non-compliance (poor engagement with the medical professionals, social/behavioural issues).

Of the 5 compliant patients, 80%(n=4) responded well with improved symptoms and blood results. Only one (20%) did not respond, He was transitioned to the adult services on UPA and after 6months of treatment needed to switch to Risankizumab due to primary non-respondent (this patient had perianal Crohn's disease and severe anxiety).

UPA shows promising results but further national and international studies are needed to prove the results found in our centre.

Table 1 Demographic details of the patient's cohort treated with Upadacitinib.

Case	Gender	**Type of IBD**	**Age at diagnosis**	**Age at starting UPA**	**Disease Duration**	**Duration of therapy**	**Previous Biologics**	**Outcome**
1	Female	Crohn's non-structuring B1, ileocolic L3	6.5ys	16.6ys	10.5ys	16ms	3	Responded
2	male	Crohn's perianal fistulating	10.5ys	14.8ys	4.5ys	Non-compliant	2	Non-compliant
3	male	Crohn's perianal Fistulating +L2 colonic	10ys	18ys	8ys	6ms	3	Primary non-respondent/ Transitioned
4	male	Crohn's non-structuring B1, ileocolic L3	9ys	15.5ys	7ys	8ms	2	Responded/ Transitioned
5	Female	Crohn's non-structuring B1, colonic L2	13ys	17ys	4ys	8ms	2	Responded /Transitioned
6	Female	Crohn's non-structuring B1, ileocolic L3	10ys	14.5ys	4ys	7ms	3	Responded

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OC35

Descriptive analysis of methotrexate in the treatment of paediatric inflammatory bowel disease: a single centre study

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Methotrexate is an immunomodulator used paediatric IBD. It is typically second line to thiopurines. This study aimed to describe reasons for methotrexate use and outcomes of methotrexate treatment at a tertiary referral centre.

A retrospective analysis of medical records from IBD diagnosis until October 2024 was performed for all paediatric IBD patients currently using methotrexate. Demographics, diagnosis, use of thiopurine and biologic medication, rationale for methotrexate use, tolerance, and response to methotrexate treatment based on serum inflammatory markers, faecal calprotectin and endoscopic re-assessment were analysed.
27 patients were identified (see table 1).
Table 1. Patients taking methotrexate

M:F	14:13	
Mean age at diagnosis (years)	10	
Mean age when starting methotrexate (years)	12	
IBD diagnosis		
Crohn's	24	89%
Ulcerative colitis	1	4%
IBDU	2	7%
VEOIBD ≤ 6 years at diagnosis	7	26%

22/27 (81%) used a thiopurine prior to starting methotrexate. The reasons for switch to methotrexate were failure of disease control (6/22, 27%) or adverse effect of thiopurines (17/22, 77%), including pancreatitis (9/22 41%), myeloid suppression (4/22, 18%), out-of-range thiopurine metabolites (3/22, 14%) and hepatotoxicity (2/22, 9%).

5/27 (19%) started methotrexate as their first immunomodulator. The reasons for this decision were low TPMT levels (2/5, 40%), rheumatological co-pathology (2/5, 40%), and parental preference (1/5, 20%).

21/27 (78%) also use biologic therapy, with 15/27 (56%) having tried more than one biologic.

26/27 patients received methotrexate subcutaneously. This was tolerated in 19/26 (73%), and the remaining 7/26 cases have been switched to oral; 6/7 (86%) for nausea/vomiting, 2/7 (29%) for abnormal LFTs, 1/7 (14%) for redness at injection site, and 1/7 (14%) for psychological disturbance. These seven patients continue to tolerate oral methotrexate.

Serum inflammatory markers (CRP, ESR, orosomucoid) were compared prior to methotrexate, at three months and six months. There was an improvement in at least two out of the three inflammatory markers at three months (17/23, 74%) and six months (13/22, 59%). Faecal calprotectin was compared prior to methotrexate and at 3–12 months. Improvement was seen in 9/12 (75%); however, it was not possible to identify calprotectin results in an appropriate time frame in 15/27 cases (56%). Of note, two patients were switched to methotrexate due to severe active Crohn's disease on their fourth biologic, prior to consideration of a JAK inhibitor or alternative biologic, and have had improvement in clinical symptoms and inflammatory markers (not yet endoscopically reevaluated).

6/27 patients (22%) had endoscopic reassessment within 12 months of starting methotrexate. Of these, 5/6 had significant active disease.

Limitations are the retrospective nature and small sample size of the study. 5/6 who were endoscopically reassessed had ongoing active disease. They may have been selected for reassessment due to ongoing symptoms, potentially biasing findings.

In summary, methotrexate is generally well tolerated and is a viable alternative to thiopurines. Switch from subcutaneous to oral methotrexate can alleviate associated nausea and vomiting. Switch of immunomodulator may be beneficial in patients who have failed multiple biologics with azathioprine. Endoscopic re-evaluation is important for full assessment of response to treatment.

OC36

Standardising Infliximab Administration: A Comprehensive Care Plan to Improve Safety, Efficiency, and Quality of Care

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Infliximab, a TNF-α inhibiting monoclonal antibody, is widely used to treat various gastrointestinal (GI) and extra-GI inflammatory diseases by targeting core inflammatory pathways. However, due to its potential side effects and risk of infusion reactions, the current guidelines recommend comprehensive pre-administration screening, ongoing monitoring, and clear multidisciplinary communication and documentation. Current workflows often rely on prescribers or specialists to manually enter orders based on varying trust-specific checklists, leading to inconsistencies and inefficiencies.

To address these challenges, we propose an order set in the form of a care plan to streamline and standardise the electronic prescribing process, enhancing efficiency and minimising errors. This care plan, developed in collaboration with the electronic prescribing and medicines administration (EPMA) team, is divided into two sections: (1) *Pre-infusion screening*, covering necessary blood tests and chest X-rays for infection screening and administration of vaccines where appropriate, and (2) *Infusion monitoring and safety*, which includes essential monitoring blood tests, safety medication, and clear documentation and equipment for nursing staff.

Analysis of DATIX data across multiple sites at a major trust revealed 37 incidents, with 10 attributed to administration errors and at least 4 due to communication and scheduling issues. Implementation of this care plan aims to:
- Reduce Medication Error: using standardised steps to minimise human error and support adherence to clinical guidelines.
- Improve Efficiency: due to the streamlined process, it prevents delays in prescribing. It also facilitates communication among the multidisciplinary team.
- Enhance Safety: by ensuring structured screening and prompt response to adverse events with medications on standby.
- Saves time: by expediting the electronic prescribing process, allowing staff to allocate more time to other tasks
- Elevates Quality of Care: by providing consistency and reliability in treatment administration.

By integrating this care plan, we aim to enhance patient safety, optimise resource use, and improve the overall quality of care for patients receiving infliximab.

OC37

Developing the Advanced Clinical Practitioner role in paediatric gastroenterology

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Introduction

The role of the Advanced Clinical Practitioner (ACP) is becoming more frequent, particularly at Sheffield Children's Hospital NHS Foundation Trust where new roles have been developed in Haematology/Oncology, PICU, surgery and ophthalmology.
The role of the ACP in gastroenterology is the first of its kind at SCH and has been in development for nearly 2 years, commencing in January 2022. Although these types of roles can offer a wide scope of practice, it is important to recognise the difficulties establishing and developing them, however once done can lead to enhanced patient care, ability to progress the speciality and offer role fulfilment.

Early stages of the role

At the beginning of this role there were no defined objectives, job plan or targeted training – which led to confusion between the ACP and other members of the team. This resulted in a difficult introduction period followed by a pressurised clinical workload which continued for 4 months until it was no longer sustainable and clearly not working in its current format. Therefore, changes were made moving forward;

Changes made to role after review:
• *Specific study days (conferences, study days related to area of practice, clinical skills training)*
• *Allocated clinic and theatre time (discussion about running ACP led clinic, possible endoscopy practice in future)*
• *More active responsibility in the team (prescribing is a big element of this, particularly homecare prescriptions due to staff unavailability)*
• *Research involvement and audits (cross centre study completed, ongoing research project involvement)*
• *Education (conference posters, participating in departmental teaching sessions, teaching junior members of staff)*
Ideas for future role development:
• *ACP follow up clinic for discharges post-acute admission; to promote* and ensure continued progress of health for patients following difficult *inpatient stay.*
• Work more closely with organisations such as BSPGHAN to create ACP *working group and deliver targeted training and learning pathways.*
• Continue research projects, especially leading on a research question of *personal interest.*
• *Upper GI endoscopy; aiming to achieve competencies in upper GI endoscopy and assist other specialities in procedures or lead own list (currently ongoing)*
• *Develop more specialty specific practice to make the role more unique, for example wireless capsule reading, PH impedance studies and* reading of results.

Conclusion

- Over a further 12 months the role successfully developed into something beneficial for the ACP and the service. Ensuring continual development is now a central focus as retrospectively more emphasis and a targeted induction/teaching/role development package would have been beneficial, with its importance being demonstrated in the early days however due to this being a new role there was a constant learning curve.
- Once recognised and acted upon the role flourished and the ACP is considered an integral part of the team and positively involved in different aspects of the service.

OC38

IgG4-Related Disease With Gastrointestinal Involvement: A rare presentation in children

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Sheffield Children's Hospital

Immunoglobulin G4-related disease (IgG4-RD) is a systemic fibro-inflammatory condition with an unclear pathophysiological mechanism affecting different parts of the body. If untreated, the disease can lead to fibrosis and irreversible organ damage.

14 year old girl, previously fit and well was transferred from local hospital with abdominal pain, temperatures, diarrhoea, weight loss (5-6kg) over 2 months. Her general and systemic examination was essentially normal. She had raised inflammatory markers with a C-reactive protein of 182 mg/Land erthyrocyte sedimentation rate of 30mm/hr. Her liver function tests were normal with albumin being lowest at 27g/. She was empirically treated with broad spectrum antibiotics.

MRI of abdomen revealed significant omental thickening with ascites. Tuberculosis and malignancy were ruled out. Initial ascitic tap was normal and histopathology – was suggestive of omental panniculitis with predominant eosinophilic cells. Initial test for autoimmune disease were negative. Subsequent MDTs identified the need to repeat the biopsy and ascitic tap (both diagnostic and therapeutic) with upper GI endoscopy, capsule study and a Bone marrow. Budd Chiari was ruled out by venography as there was high flow in hepatic vein confluence which was probably due to the pressure effect of ascites.

Repeat ascitic fluid SAAG was 0.4 g/dl confirming an exudate , cell count could not be done as sample clotted and histopathology was suggestive of sclerosis /fibrosis with plasma cells .IgG4 staining was 40% and serum IgG4 was normal. Stains for IMT (inflammatory myofibroblastic tumour) were negative and genetics awaited for IMT and familial mediterranean fever gene.

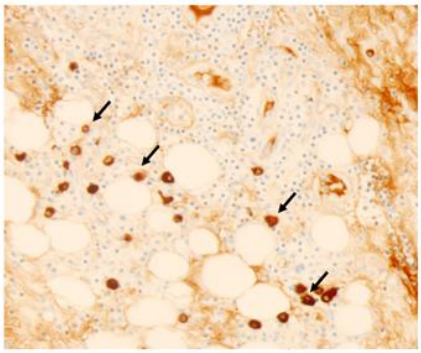
She improved clinically with resolution of inflammatory markers at the time of second biopsy. She is asymptomatic at present. After extensive review of literature and discussion with a quaternary centre she was diagnosed as IgG4 related disease. In this case her condition resolved spontaneously without the need for immunosuppression.

IgG4-related disease (IgG4-RD) is a chronic, systemic, and autoinflammatory disease mediated by the immune system.

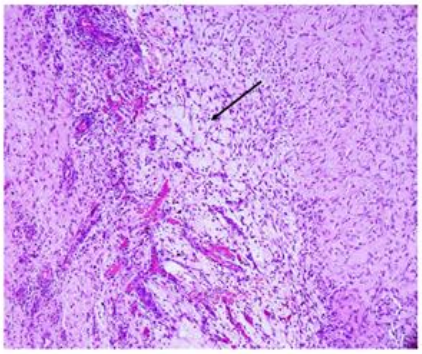
Gastrointestinal presentation generally involves pancreas. In this case it was an isolated involvement of omentum with a pleural effusion.

This presentation is rare. There are no reported cases of omental involvement as presentation of this disease in children.

Groups of IGG4 + cells



Foamy macrophages amongst inflammation



OC39

Gastrointestinal bleeding related to mTOR inhibitor: a rare paediatric case

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Sheffield Children's Hospital

Gastric antral vascular ectasia (GAVE) associated with mTOR inhibitors has been reported in adult literature(1,2), but to our knowledge there is no published data of targeted drug therapies associated with severe gastrointestinal haemorrhage in paediatric patients. GAVE itself is considered a very rare cause of gastrointestinal bleeding in children.

We present a case of Everolimus- associated gastrointestinal bleeding in a paediatric patient.

A 15-year-old boy with a background of Tuberous sclerosis, cardiac rhabdomyoma, seizures and recurrent fractures presented with a life-threatening gastrointestinal bleed. He had been treated with Everolimus since 2014 as part of drug trial for cardiac rhabdomyoma.

Whilst in hospital for a lower respiratory tract infection, he developed hematemesis and required blood transfusion (Hb dropped from 92 to 81 g/L). He underwent OGD with banding of vascular lesions in his stomach by adult gastroenterologists. He developed further melena, hence he was transferred to our Unit where he was treated with octreotide infusion. In view of on-going melena and oxygen requirement, he underwent endoscopic reassessment that revealed generalised oozing in gastric antrum (Figure 1). Haemospray was applied which stopped the oozing (Figure 2). He dropped his haemoglobin again to 75g/L (from 88 g/L) with worsening of his respiratory and neurological status. He was transferred to high dependency unit and received another blood transfusion and parenteral nutrition. At this point, endoscopic reassessment under GA wasn't deemed safe. Octreotide infusion (5mcgrams/kg/minute) continued. Computed Tomography scan of the abdomen ruled out vascular anomalies. Detailed review of his medical background led to literature search on Everolimus and its association with gastrointestinal bleeding. Everolimus was withheld. As he had tolerated the mTOR inhibitor for 10 years, the neurology team were keen to restart it. 72 hours after restarting Everolimus, he dropped his haemoglobin from 93 to 75g/l. Following MDT discussion, everolimus was discontinued.

Endoscopic re-evaluation revealed vascular lesions and two band ligators (7mm) were applied distally to proximally in gastric antrum (Figure 3). Subsequently, he was successfully weaned off octreotide and was re-established on oral diet without any further gastrointestinal bleeding in the next 6 months.

We successfully treated GAVE with band ligation in a paediatric patient who was treated with mTOR inhibitor. GAVE is considered to be associated to mechanical stress caused by altered gastric motility and gastric dysfunction that leads to submucosal fibromuscular hyperplasia and dilation of mucosal capillaries. Everolimus increases the gastric motility which may contribute to the development of GAVE. We aim to raise the awareness of this rare adverse effect which can be a challenging cause of severe gastrointestinal haemorrhage in children.

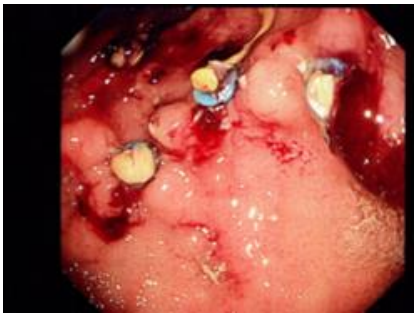

Figure 1:Endoscopy showing previous bands and oozing in the antrum

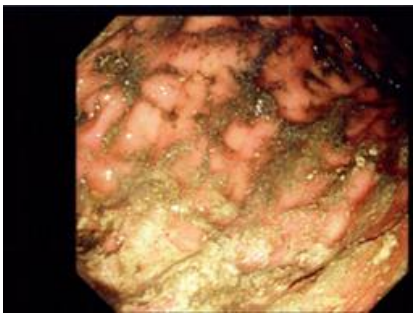

Figure 2: Application of haemospray

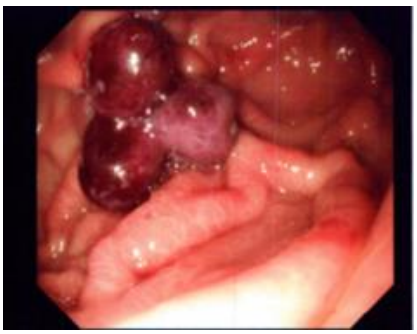

Figure 3: Application of bands to the vascular lesions in the stomach

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OC40

Acute gastroparesis in association with acute pancreatitis in an adolescent

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Despite the increasing prevalence and healthcare costs, many cases of acute paediatric gastroparesis (GP) remain idiopathic.[1-] In adults, GP is often associated with diabetes or post-surgical conditions, while approximately 70% of paediatric GP cases are idiopathic and 12% are post-surgical [2] Notably, around 7% of GP patients have a history of pancreatitis, and 44% of chronic pancreatitis patients experience GP.[3-] Research by Parkman et al. suggests that patients with both conditions experience worse symptoms, have a higher likelihood of requiring total parenteral nutrition, and report lower quality of life compared to those with GP alone.[4]

The overlap of common symptoms such as abdominal pain, nausea, and vomiting, makes diagnosing acute GP particularly challenging.

GP is characterized by delayed gastric emptying and ranges from mild discomfort to severe complications, including dehydration and malnutrition. Diagnosis is difficult due to lack of standardized criteria. While paediatric GP data is limited, hospitalization rates have significantly risen over the past decade, impacting quality of life and increasing morbidity and mortality. [5]

A previously healthy child presented with a 12-hour history of severe epigastric pain, recurrent vomiting and fever. Clinical and biochemical assessments indicated acute pancreatitis showing elevated inflammatory markers (CRP 32mg/L) and significantly raised amylase of 1786 iu/L. Initial nasogastric decompression yielded over3 litres of dark green bilious aspirates

A Computer Tomography (CT) imaging was organised to exclude other intra-abdominal pathologies, confirm the diagnosis of pancreatitis and evaluate for potential gallbladder stones. This showed a grossly distended stomach with a distended, fluid filled second part of the duodenum, a narrow third part, and a collapsed remainder of the small and large bowel. Free fluid surrounding the pancreas suggested acute pancreatitis with acute gastroparesis. Subsequent magnetic resonance imaging showed significant inflammatory changes around the pancreatic head and a distended stomach. No calculi were observed in the common bile duct.

The patient was treated conservatively, with nasogastric decompression, broad spectrum antibiotics, intravenous fluids and a proton pump inhibitor. In view of persistent large gastric aspirates, Polymeric feeds (Nutri Peptisob Energy) were started via a naso-jejunal tube and gradually reduced once oral intake was tolerated. Possible causes, including infection and autoimmune pancreatitis were excluded with negative blood and stool cultures, viral panel, IgG serology and normal genetic screening for pancreatitis (PRSS1, SPINK1, CFTR, CELA3B).

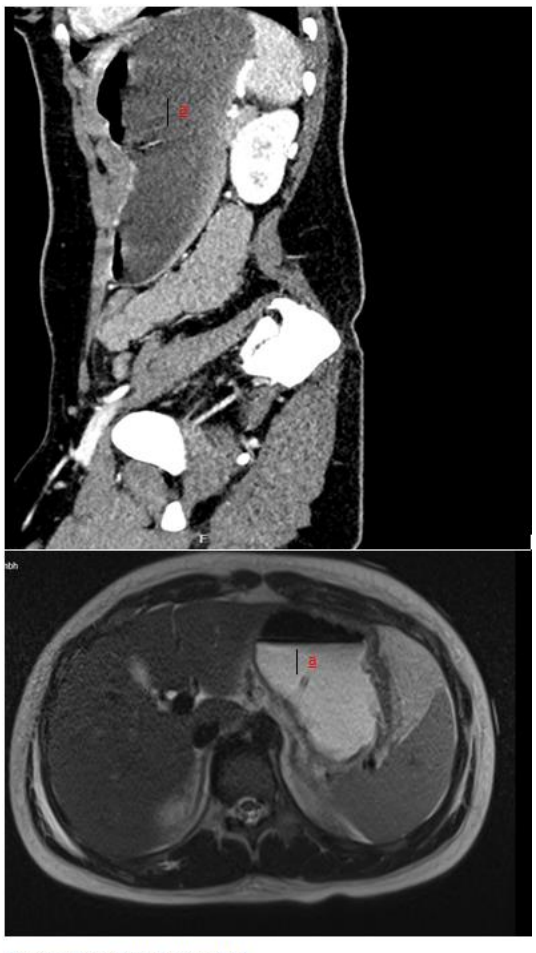

Figure 1 – Grossly distended stomach (a)

Upon discharge from Hospital, patient was tolerating normal diet and had normal faecal elastase.

At her most recent follow-up, she was thriving, well and had no symptoms suggestive of gastroparesis or pancreatic insufficiency.

This case highlights the need for increased awareness and discussion of such atypical presentations. While acute pancreatitis elevates the risk of developing gastroparesis and increases hospitalization rates[6], there is no current consensus on the optimal management strategy in acute settings. This emphasizes the necessity for further research into paediatric GP, particularly its connection to pancreatitis, as cases grow in both frequency and complexity.

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OC41

Management strategies for recurrent idiopathic pneumatosis coli.

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We present the case of a 17-year-old boy with a known case of: Alexander disease, Haemophilia A, epilepsy and Gastrojejunostomy (GJ) feeding. He was admitted to the Emergency Department (ED) with a two-day history of worsening abdominal distention and left sided abdominal tenderness. Initial laboratory tests revealed a raised White Cell Count and CRP (64). Abdominal X-ray (AXR) demonstrated significant large bowel distension *(Figure 1)* and a subsequent Computed Tomography (CT) indicated distended large bowel loops with a prominent caecum, pneumatosis and small pneumoperitonem (*Figure 2*). Importantly, the degree of pneumoperitoneum and clinical presentation did not indicate bowel perforation.

The patient was managed conservatively with broad-spectrum antibiotics, gastric decompression, and placement of a flatus tube. The jejunal tube was placed on free drainage. The patient had experienced a similar episode several months prior, which was also treated conservatively.

Over the following 5 days, his abdominal distension and gastric aspirates decreased. A Peripherally Inserted Central Catheter (PICC) line was inserted, and feeds were gradually reintroduced.

Eight weeks later, the patient returned to the ED with recurrent vomiting and worsening abdominal distension. Repeat AXR and CT imaging revealed worsening large bowel dilation, persistent right colonic pneumatosis, pneumoperitoneum, and a small amount of free fluid.

Multidisciplinary team discussions were held both regionally and at super specialist level to formulate the best mode of treatment for this complex patient. It was agreed to continue with a conservative approach.

This was well tolerated, and the patient's condition improved. The elemental feed was reintroduced, and progressively increased.

On all occasions, the patient's condition was successfully managed conservatively.

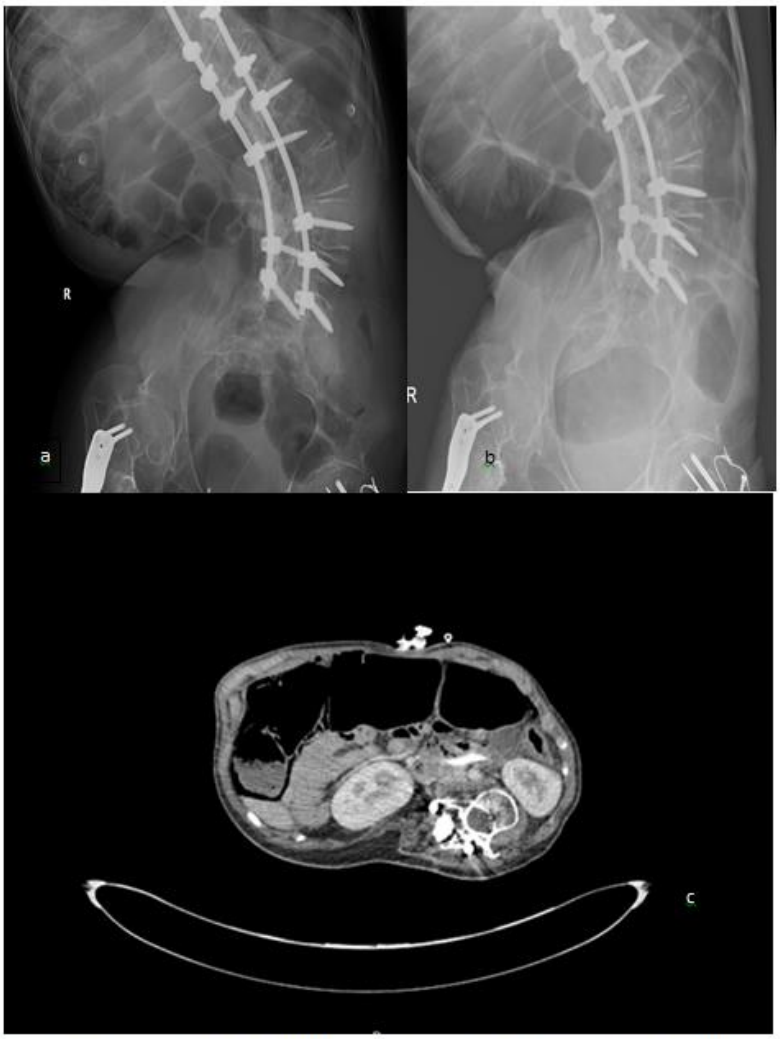

Figure 1- AXR – large bowel distension – a) First b) Second presentation c) CT findings of Pneumatosis intestinalis

This case highlights the increasing incidence of similar presentations, and the complexities involved in treating patients with neurological impairment and gastric dysmotility. Conservative management of PC should be considered in paediatric patients with neurological disabilities, intestinal dysmotility, and those dependent on long-term feeding tubes. Elective surgical treatment may be warranted in selected cases for recurrent episodes of PC who fail conservative treatment.[1-2]

Given the variability in the literature regarding the management of PC, and the recurrent nature of the condition in this cohort of patients, the development of clinical practice guidelines at regional, national, and international levels should be considered to ensure a standardized, conservative approach to care.

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OC42

Classical homocystinuria (HCU) presenting with complications of chronic pancreatitis

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Classical Homocystinuria (HCU) is a rare inherited disorder of metabolism caused by deficiency in cystathionine-β-synthase (CBS), leading to accumulation of homocysteine and its precursor methionine. Untreated, there is a risk of thromboembolism as well as complications involving eye, skeleton and brain. We present a case, presenting with complications of chronic pancreatitis, subsequently diagnosed with HCU.

A 14-year-old male had a 9 month history of episodic abdominal pain, diarrhoea, vomiting and weight loss. Initial investigations revealed normal Coeliac serology and faecal calprotectin and a raised serum amylase 162 U/L. He developed shortness of breath and a CT scan confirmed a large right sided pleural effusion requiring chest drain insertion. Pleural amylase was raised; 2817 u/L and a pancreatic-pleural fistula secondary to acute on chronic pancreatitis was diagnosed. CT reported pancreatic inflammation, thickened small bowel loops; ischaemic wedge infarct of the liver; portal vein thrombosis and multiple small stones in the gall bladder. He was managed conservatively with total parenteral nutrition (TPN).

Labs revealed raised alanine transferase (ALT) 76 U/l (10 – 50 U/l), hypoalbuminaemia 15 g/L (35 – 50 g/L), low fat soluble vitamin levels A, D, E and a coagulopathy; prothrombin time 21 s (normal range 10.0 – 13.2) triggering metabolic investigations. Blood amino acid profile identified elevated plasma methionine (>2000 umol/L) and total homocysteine (tHcy)(>200 umol/L). Classical HCU was diagnosed. Urgent action to reduce TPN protein, reducing the risk of cerebral oedema and thromboembolism, using methionine free PN, proved challenging. IV pyridoxine, folate and anticoagulation were started. Small bowel oedema persisted and Creon (faecal elastase < 15 ug/g stool (> 200 ug/g stool) was started. Nasogastric feeds led to an increase in gastrointestinal symptoms and raised serum lipase. However, nasojejunal feeding enabled transition from TPN to enteral feeding. Plasma methionine (17umol/l) and tHcy normalised (3.4umol/L) over 7 weeks. He was discharged home on full nasojejunal feeds and recently has re-establsihed oral eating.

Chronic pancreatitis is a rare complication of HCU, possibly caused by thromboembolic events in the vasculature of the pancreas. Only a few cases have been reported in the literature.

OC43

Follow-up practices for paediatric patients with intestinal failure graduating off home parenteral nutrition

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Tertiary Gastroenterology departments oversee the nutritional care of patients with Intestinal Failure requiring Home Parenteral Nutrition (PN) with support from district general hospitals. The South-West of England lacks a guideline for nutritional monitoring once children discontinue PN and establish full enteral feeds.

In order to explore the current care provision, information was sought regarding the frequency of blood monitoring, growth monitoring and paediatrician follow-up once PN was discontinued.

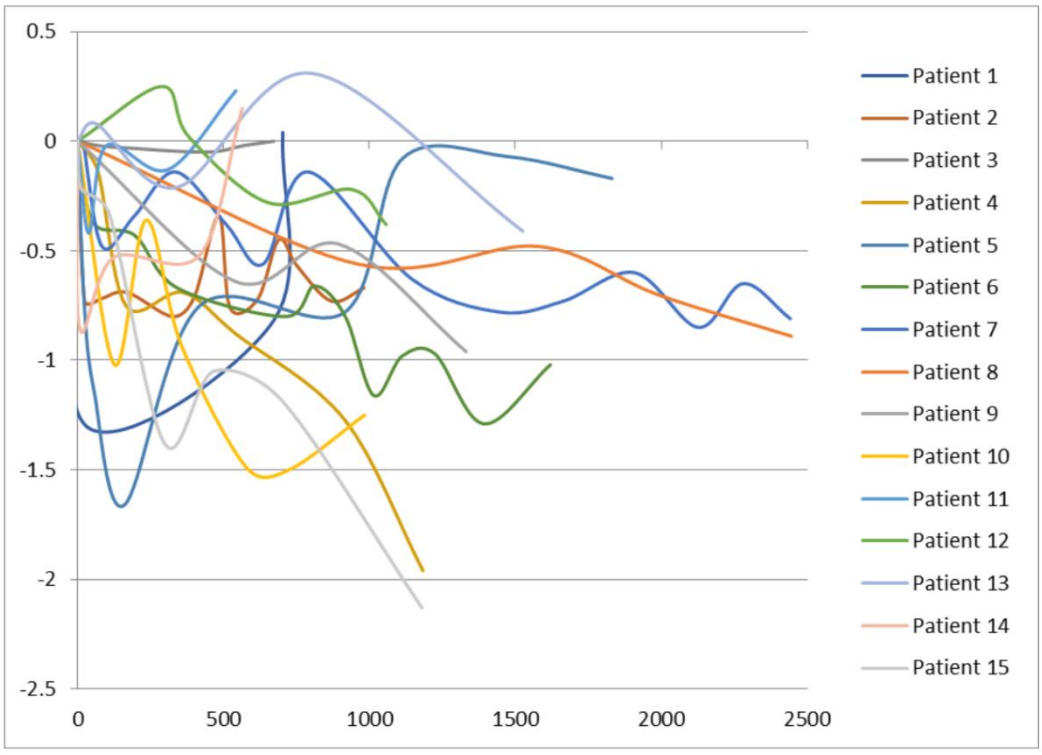
Data was obtained from 23 patients within the South-West 'Paediatric Intestinal Failure requiring Home Parenteral Nutrition' database (initiated 2015) whose PN was discontinued before February 2024. 3 patients died of unrelated comorbidities, 3 transitioned to adult services, 1 moved region and 1 established full enteral nutrition following a diagnosis of Induced Illness. The remaining 15 (9M, 6F) were identified with diagnoses including Atresia, Gastroschisis, Volvulus, Necrotising Enterocolitis and Pseudoobstruction. Electronic records, laboratory results and correspondences with local hospitals identified all outpatient follow-up and blood monitoring led by gastroenterology consultants.

Patients experienced 2.0 reviews per year of which 1.4 were face to face. At some point during their follow-up, half of the patients experienced a gap exceeding 12 months with no clinical review or monitoring. The median frequency for height and weight recording was 1.2 and 1.4 times per year.

Assessment of change in Z scores from a baseline (See Graph 1) set at the point of discontinuing PN demonstrated a negative trend with the majority of patients falling weight centiles over time. Those falling by 2SD were highlighted to their local hospital for additional dietetic support.

Nutritional blood checks were performed less than once per year, with several nutrients and vitamins rarely checked. 11/15 patients were noted to be vitamin deficient at some point in their follow-up. Many patients required supplementation with Iron, Folate, B12 and Vitamin D during the follow-up period.

Z score vs Time since stopping PN (Graph 1)



The data highlights that patients with Intestinal Failure discontinuing PN continue to require close nutritional monitoring as many develop deficiencies requiring supplementation, only detectable with proactive monitoring. Although most experienced frequent reviews and growth measurements, the findings highlight that the point of discontinuing PN may represent the Childs peak nutritional state with a subsequent decline without careful follow-up. Although COVID19 may have interrupted normal service delivery, a clear care plan needs to be developed to inform families and professionals on optimal nutritional monitoring once successfully graduated from PN.

OC44

Assessing bone health in paediatric intestinal failure patients on home parenteral nutrition: A tertiary centre experience

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Metabolic Bone Disease (MBD) is a significant complication in children with intestinal failure (IF) on home parenteral nutrition (HPN). Clinical symptoms may not be informative; MBD can be asymptomatic or present with fractures. Routine monitoring includes biochemical markers such as serum vitamin D, alkaline phosphatase (ALP), plasma phosphate, calcium, and parathyroid hormone (PTH) levels, while DEXA remains the gold standard for assessing osteoporosis.

This study aimed to assess bone health in children on HPN and identify markers for early intervention. A retrospective review of patients' clinical records concentrated on DEXA scans, peripheral quantitative CT (pQTC) measurements, and biochemical parameters, using the nearest results to the DEXA scans. Patients with kidney disease and secondary hyperparathyroidism were excluded from the analysis.

Out of 36 children, 24 had at least one reported DEXA scan (>5 years of age) and were included in the analysis. The median age was 12 years (5–18 years), 45.8% male. The mean BMI z score was –0.39± 0.96 (p=0.29) and height z score –1.96±1.2 (p<0.001).

20.8% (5/24) had congenital enteropathies, 75% (18/24) had short bowel syndrome, and 8.3% (2/24) had chronic intestinal pseudo-obstruction. Median HPN duration was 10.53 years (2.94–17.65 years). 44.4% (12/24) had at least one DEXA parameter z score ≤ -2, indicating low bone mineral density (BMD). 13.6% (3/22) had temporarily low mineralization, and 31.8% (7/22) showed temporary changes in spinal morphology due to osteopenia. One patient had fractures. Follow-up scans showed persistent low BMD in 75% (6/8), though bone mineralization and spinal changes improved with regular biochemical monitoring and supplementation.

The most consistently reported parameters are shown in Table I.

Table I. DEXA metrics in paediatric home PN patients

	Mean z score (n=24)	one tail t test (p value)
BMD whole body less head for age	-4.00 (SD= 9.1)	-2.02 (p=0.028)
Bone mineral apparent density spine (BMAD)	-0.52 (SD= 1.43)	-1.72 (p=0.049)
pQTC4%		
Trabecular	0.14 (SD=1.6)	0.41 (p=0.34)
Cortical and Trabecular	0.53 (SD=1.55)	1.58 (p=0.06)

Only 3/24 patients had elevated PTH levels for >2 months. Adjustments were made to the calcium/phosphate ratio in the PN, or additional calcium supplementation was provided. Vitamin D was routinely supplemented to maintain normal levels (mean 25-OHD3 69.7±23.3).

All patients with abnormal scans were reviewed by the paediatric endocrine team and 2/24 have been diagnosed with osteoporosis and started on treatment.

Children with intestinal failure are at high risk for bone complications, including fractures, even with regular monitoring and supplementation.

Low bone mineral density, once diagnosed, has been persistent in our population, though mineralization can improve with vitamin D and calcium supplementation.

BMD and BMAD z scores were significantly lower in our cohort, while 4% pQCT cortical/trabecular and cortical z scores did not differ to the general population. DEXA metrics are height-dependent, but pQCT is less affected by patient height, providing a potentially more accurate measure of bone mineral density.

Regular DEXA and pQCT scans should be standardized for paediatric intestinal failure patients on HPN, especially those with significant BMD reduction or fracture history.

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OC45

Syndromic congenital sodium diarrhoea – phenotypic variability in three cousins

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Congenital Sodium Diarrhoea (CSD) is a rare, genetical form of intractable diarrhoea with high faecal sodium loss. Symptoms start in utero; life-threatening dehydration, electrolyte abnormalities and metabolic acidosis occur if untreated. Syndromic CSD caused by SPINT2[1] mutations is associated with choanal/intestinal atresias, corneal erosions and tufting enteropathy. We present three male infant Kuwaiti cousins with syndromic CSD and variable clinical presentation.

Case 1 born at term, birth weight (BW) 3.6kg, presented with diarrhoea and severe dehydration on day 20. Pregnancy was uneventful (absent polyhydramnios). On transfer at 11 months he was on minimal enteral feeds, suboptimal parenteral nutrition (PN) and severely malnourished (3.9kg, z-7.68)). Urine sodium was low (< 5 mmol/L), faecal sodium high (113 mmol/L). Diarrhoea continued when fasting. OGD revealed duodenitis with tufting-like changes of surface enterocytes (EpCAM retained). Genetic testing revealed a homozygous SPINT2 gene variant, confirming CSD. After stabilization with PN and correction of fluid/electrolyte abnormalities he achieved slow catch-up growth (6.84 Kg, -4.10) and was repatriated on PN (160ml/Kg, sodium 10 mmol/Kg) with minimal oral diet aged 16 months.

Case 2 born at term (BW 3kg), passed meconium, had formed stools initially but presented dehydrated with diarrhoea on day 10 following uneventful pregnancy (no polyhydramnios). On transfer aged 3 months his weight had dropped to 2.85kg (z- 5.00). PN was started; feeds temporarily discontinued. Diarrhoea continued. Stool sodium was high (127 mmol/ml). OGD demonstrated tufting enteropathy (EpCAM preserved). Genetic testing is pending but SPINT2 mutation likely; CSD diagnosed clinically. At present he is 10 months old with good weight gain (8.47kg, 25th centile). He remains on PN (210ml/Kg, sodium 18 mmol/Kg) awaiting home PN.

Case 3 born prematurely (32/40), BW 1.8kg (50th centile corrected, z= 0.00), antenatal polyhydramnios, developed diarrhoea soon after birth, which continued when fasted. PN was started (severe dehydration, electrolyte disturbance). Choanal atresia was repaired in the first week. Genetic testing identified a homozygous missense variant in SPINT 2 gene confirming CSD. When transferred he was malnourished (2.35 Kg, z- 7/16). PN fluids + electrolytes were optimised (120 ml/Kg, sodium 10 mmol/l), macronutrients gradually increased (high refeeding risk) and stool losses replaced ml for ml with 0.9% NaCl + KCL 10 mmol/500ml (stool sodium 120mmol/L). Endoscopic assessment is pending.

Each case had similar dysmorphic features but no eye problems.

Syndromic CSD is clinically heterogeneous. Only 1/3 in our series had antenatal polyhydramnios, presented with severe diarrhoea on day 1 of life and had choanal atresia. 2/3 cases developed diarrhoea later (day 10 and day 20 respectively), although this may not be accurate as stool may have been mistaken for urine.

Early initiation of PN and replacement of fluid and electrolyte losses is essential to avoid malnutrition and life-threatening fluid/electrolyte imbalance. Antenatal polyhydramnios and/or bowel dilatation is not always seen. Clinical suspicion after birth should be high. A stool sample for electrolytes can be easily obtained though insertion of a small-bore nasogastric tube into the rectum to differentiate urine from sodium rich stool[2].

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OC46

First Steps: Learning from an Inaugural Parenteral Nutrition (PN) Family Day

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Our nutrition support team (NST) ran an inaugural Home Parenteral Nutrition (HPN) Family Day in October 2024, for children and young people receiving HPN and their families, including siblings. Parallel sessions included organised entertainment (disco party, balloon modelling, magic show) for the children and young people, running alongside educational events for parents and caregivers. There were presentations from the clinical psychologist, clinical nurse specialists, along with input from patients and carer representatives to share their experiences. The event concluded with a panel discussion and networking event alongside a teddy making workshop.

We were keen for feedback from both the families and the healthcare professionals involved. To achieve this we devised a questionnaire. Participation was voluntary and the information collected was anonymous. Questions were a combination of binary questions and free text opportunities.

There are 14 children and young people under the care of the NST on HPN. 10/14 families attended the Family Day. 9 parents & 1 young person competed the family questionnaire.

100% of families reported the day was “extremely useful” and 100% reported that the level of information was “just right.” The feedback for the panel discussion: 70% of respondents reported that it was “extremely useful,” and 30% indicated it was “somewhat useful.” 80% of respondents indicated they “very much enjoyed” the children’s activities. 90% of respondents indicated it was “extremely useful” to meet other families of children on HPN. We asked families what they liked about the day. Comments included “meeting others in the same situation,” “feeling validated for all the feelings we have, dealing with a chronic illness and home PN,” “hearing about tips and tricks.” “What the gastro team wants the future to look like for their patients.” We asked families what we could improve for any future family days and comments included “next time parents and older children could have input into the agenda,” “maybe an ice breaker for parents to introduce themselves.” The last question asked families what issues matter most to them.57% of families answered the lack of a specialist home HPN nurse was an key issue for them. Other feedback included, advice for swimming with central lines and families were keen for future events including informal and recreational events, suggesting various activities they would enjoy.

The family feedback reflects the fun, positive atmosphere of the day and how meaningful it was to families to meet each other and share experiences. We have presented the feedback at our multidisciplinary team meeting and look forward to sharing it with the hospital mangers. highlighting the issues that matter to the families, including the need for a dedicated HPN nurse. This inaugural event was a success and we are keen to plan similar events in the future, co-producing these with parents and young people themselves. Through these activities we build community and can better advocate for the families that use our service.

OC47

Demographics and Outcomes of Central Venous Catheter Repair in a Single Paediatric Intestinal Failure unit

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For children with intestinal failure requiring parenteral nutrition (PN), their central venous catheter (CVC) is essential, to provide nutrition and fluid for growth, development, education and often life. These catheters are at risk of infection, thrombosis and damage and frequent line replacements can lead to loss of this life-preserving access.[1] Current practice aims to preserve vascular access, of which part is repairing damaged CVCs in the first instance.[1] The aim of this study is to describe the children requiring line repairs in a single paediatric intestinal failure unit and review the outcomes following line repair.
All children registered with a single intestinal failure unit who underwent a CVC repair over a two-year period were identified (January 2021- December 2023). 9 children (7 male) underwent a total of 19 CVC repairs (range 1-5 repairs per patient). Intestinal failure was secondary to small bowel syndrome in 56%, the remainder dysmotility of varying causes. Median age at CVC repair was 6 years 1month (interquartile range (IQR) 3years 7 months to 7 years 9 months) 5 children had more than one CVC repair during the study period and 2 CVCs were repaired 3 times. At time of CVC repair 17 were on 7 nights PN with a median of 12hrs PN per night (range 11-24). The remaining child (2 CVC repairs) was on fluids only. At presentation the repaired line was the child's 5th line(median) (IQR 3-5)

Median age of CVC at repair was 253 days (IQR 188.5-377). The CVC had snapped in 5/19 cases, the remainder with visible fractures, holes, or loss of integrity. In 18 cases, the CVC was repaired by the on-call surgical registrar, the remainder by a haematology nurse practitioner. 5/19 (26%) CVC repairs were unsuccessful, requiring early (<7 days) replacement or further repair. Median CVC survival to further repair or replacement was 43 days (IQR 5-113) and from first repair to replacement 85.5 days (IQR 51.75-174). There were no central line associated blood stream infections (CLABSI)within 30 days of line repair, and only one CLABSI in a repaired line during the study period. (0.76 per 1000 catheter days).

CVC repair is an effective and safe tool in prolonging a CVC lifespan, with a low infection rate in our cohort, similar or better than published literature.[1-6] Early repair failure was relatively high in our cohort (26%) and may reflect varying levels of experience in the professionals performing the procedure. Intensive training is currently underway with a small cohort of staff with the aim of reducing the total number but increasing the experience and competence of staff undertaking CVC repair.

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OC48

Investigation of urgent reasons for central venous catheter replacements and outcomes in paediatric home parenteral nutrition patients

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Patients with intestinal failure (IF) require central venous catheters (CVC) for administration of long-term parenteral nutrition (PN). The presence of a CVC is a major risk factor for severe complications like central line associated bloodstream infections (CLABSIs), thrombosis and mechanical complications (occlusions, fractures or displacements) which can require an urgent CVC replacement.
The aim of this study was to investigate urgent reasons for CVC replacement and outcomes in paediatric Home PN (HPN) patients.

Retrospective data was collected from electronic notes of patients receiving HPN from January 2022 to September 2023. From 27 patients managed with HPN during that interval (6 started during that period, one stopped PN and one transitioned to adults), 11 patients (7 females) with a median age of 12 years 1 month [IQR 7y 1m – 13y 9m] required acute line changes. 3 patients had multiple line changes during this period (two had 2 changes and one had 3 changes), so in total we describe 15 acute CVC changes. At the time of the change, the median of the PN nights was 7 nights given over a median of 12 hours [IQR 12 - 24 hours].

Indications for HPN in these patients were short bowel syndrome (4 cases), gut dysmotility (5 cases) and microvillus inclusion disease (2 cases). PN was delivered through a single lumen tunnelled CVC except for one patient who had a double lumen CVC. The CVCs were locked with Taurolock in most of the cases (except one patient with Taurolock allergy). 6 981 catheter days were evaluated during the study period. Causes for acute line changes were: CLABSIs (4 cases – 2 Candida, 2 Staphylococcus aureus) and one exit site infection, followed by mechanical complications - broken lines (4 cases), displacements (4 cases) and occlusion (1 case). The infection rate was 0.57 per 1000 catheter days. Line changes were performed by paediatric surgery in 9 cases and interventional radiology in 4 of the patients. The median time between admission and line change was 2 days [IQR 1 -3 days]. The median length of admission for line change was 3 days [IQR 3 – 7.5 days]. The length of admission increased for patients who needed a line change due to CLABSIs due to antibiotic treatment. In patients who underwent more than one line change, the change was due to mechanical complications, 3/4 displacements being seen in patients with learning difficulties.

In conclusion, although the CVC related complications have decreased over the years due to medical advances and multidisciplinary team approach, these are still happening in paediatric HPN patients. Some of the CVC replacements are done acutely and our study showed the reason for the change can be related with comorbidities (e.g. learning difficulties, severe IF with 24 hours PN dependency).

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OC49

Tacrolimus in children with autoimmune hepatitis

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The first line treatment for autoimmune hepatitis with Azathioprine and prednisolone is effective in 80-90% children. Tacrolimus is used as second or third-line agent in children with 75%- 94% attaining biochemical remission.

This is a retrospective audit on the use of tacrolimus in autoimmune hepatitis in children ≤16 years from 2010- 2024. Demographics, indications of tacrolimus use, clinical course, laboratory parameters, liver biopsies (fibrosis staged by Ishak fibrosis score), treatment response, complications and outcomes of therapy (remission, relapse or liver transplantation) were assessed. Biochemical remission was defined as alanine aminotransferase <40 IU/ml.

Tacrolimus was used as second (n=7) or third (n=4) immunosuppressant in 11 children over a time period of 14 years. Tacrolimus was started at a median age of 14.5 (IQR 13- 16) years and interval between diagnosis of autoimmune hepatitis and initiation of tacrolimus was 14 (9 -60) months. The indications for changing to tacrolimus were failure of first line therapy (n=10), complications related to previous immune suppressive therapy (n=5) and non-compliance with previous therapy (n=2). Progression of fibrosis stage was noted in 72% (n=8) at the time of initiation of tacrolimus as compared to the liver biopsy at diagnosis. Initial biochemical remission was attained in 55% (n=6) and only 27% (n=3) attained long term remission with tacrolimus therapy. The total duration of follow-up on tacrolimus was 17 (IQR 11- 32) months. Five children (45%) were listed for liver transplantation while three children were started on additional biological therapy. There were no complications from the use of tacrolimus during the study period.

Tacrolimus might be useful as an alternative immunosuppressive drug in a very select group of children with treatment failure, incomplete response, or intolerance to first-line agents. Careful selection of patients is necessary as those with advanced liver disease with initial treatment failure is unlikely to respond, with progression of disease over time necessitating addition of biologics or need for liver transplantation.

OC50

Improvements in pruritus after maralixibat treatment are associated with improved health-related quality of life for patients with cholestatic liver disease

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Alagille syndrome (ALGS) and Progressive Familial lntrahepatic Cholestasis (PFIC) are rare cholestatic liver diseases (CLD) associated with severe pruritus along with markedly reduced health related quality of life (HRQoL). Maralixibat (MRX), an oral minimally absorbed ileal bile acid transporter (IBAT) inhibitor, is approved for the treatment of cholestatic pruritus in patients with ALGS ≥2 months of age and for the treatment of PFIC ≥3 months of age in the EU, respectively. This analysis assessed whether improvements in pruritus after MRX treatment are correlated with improvements in a variety of HRQoL domains in these CLD.

The study designs of the Phase 2 randomized withdrawal period (RWD) ICONIC trial and Phase 3 randomized, double-blind, placebo-controlled (PBO) MARCH trial have been previously described. Separate retrospective analyses of pruritus and HRQoL data from the ICONIC trial in ALGS (18 weeks of open-label MRX treatment) and the MARCH trial in PFIC (26 weeks of MRX or PBO treatment) were conducted. Patients in both studies had to have moderate to severe pruritus as measured using a validated caregiver-reported Itch Reported Outcome (ltchRO) severity assessment tool (0=none to 4=very severe). HRQoL was assessed using the Pediatric Quality of Life Inventory Generic Core (PedsQL; 0-100 scale, 100=best quality of life), Physical Health (PH), Psychosocial Health (PSH), and Multidimensional Fatigue (MF) scale scores, which were collected via caregiver in both studies. In MARCH, a subset of questions from the exploratory diary questionnaire (EDQ) focused on sleep disturbance were assessed for their relationship to pruritus improvement. Spearman's (r) coefficients were determined to evaluate the relationship between pruritus improvements and HRQoL.

A total of 28 patients with ALGS from ICONIC were included with a mean± SD baseline age of 5.4 ± 4.2 years, ltchRO score of 2.9 ± 0.5, PedsQL score of 60.3 ± 16.6, PH score of 64.7 ± 20.0, PSH score of 57.6 ± 16.7, and MF score of 51.2 ± 22.6. After 18 weeks of open-label MRX treatment, there was a significant positive correlation between pruritus improvement and improvements in Peds QL (r 0.50 [p=0.007]), PSH (r 0.47 [p=0.012]), and MF (r 0.71 [p=0.0002]). In MARCH, 55 patients from the AII-PFIC cohort (29 MRX, 26 PBO) were included in the analysis with a mean± SD baseline age of 4.9 ± 4.1 years, ltchRO score of 2.9 ± 0.9, PedsQL score of 55.8 ± 19.0, PH score of 61.5 ± 22.4, PSH score of 52.0 ± 20.2, MF score of 57.8 ± 20.4 and EDQ sleep disturbance score of 3.7 ± 0.8. Among MRX-treated participants after 26 weeks of treatment, there was a significant positive correlation between pruritus improvement and improvements in Peds QL (r 0.53 [p=0.003]), PH (r 0.50 [p=0.006]), PSH (r 0.47 [p=0.01]), and sleep (r 0.96 [p<0.0001]). In the PBO-treated participants, improvements in pruritus were significantly correlated with improvements in PSH (r 0.41 [p=0.039]) and sleep (r 0.88 [p<0.0001]).

These data further illustrate the robustness of MRX's impact on the relationship between pruritus improvement and HRQoL across multiple domains. Irrespective of the CLD studied, improvements in pruritus after MRX treatment were strongly associated with improvements in HRQoL.

OC51

Hepatopulmonary Syndrome and Portopulmonary Hypertension in children with portal hypertension: a single-centre experience

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Hepatopulmonary syndrome (HPS) and Portopulmonary hypertension (PoPH) are both pulmonary complications of advanced liver disease associated with high mortality and poor prognosis(1). HPS is arterial hypoxaemia (PaO_2 <80kPa) with an increased alveolar-arterial oxygen partial pressure difference ($AaPO_2$ >15) on a background of liver disease(2)(3)(4) which is a consequence of pulmonary vasodilatation. PoPH is characterised by pulmonary arterial hypertension with co-existing portal hypertension(5)(6).
The aim of this 20-year retrospective review (2003-2023) is to report our single centre experience to further delineate the clinical presentation, diagnosis, and treatment for children with HPS and PoPH. We identified patients through the database and collected data through evaluation of electronic case records.

We identified 7 children (4 female and 3 males; age range 10 months-13 years) with a diagnosis of HPS. Their underlying aetiologies comprised of biliary atresia (2), portal cavernoma (2), veno-occlusive disease (1), non-cirrhotic portal hypertension (1) and peroxisomal biogenesis disorder (1). 4/7 patients presented with dyspnoea and 4/7 showed clinical signs of cyanosis and clubbing at presentation. Oxygen saturations (SpO_2) in air ranged from 82-89%. 3/7 patients had congenital cardiac disease. Of the non-cardiac patients, only 1 patient had a normal Chest X-Ray, the others demonstrated coarse bronchovascular markings. For diagnostic confirmation, 6 patients had contrast enhanced echocardiography, which were all positive and demonstrated evidence of shunting. Macroaggregated albumin (MAA) perfusion scanning identified abnormal extra pulmonary shunt fractions (25-73%) in all patients.

3 cardiac catheter reports were available for analysis. They demonstrated normal pulmonary artery pressures and pulmonary vascular resistance however, showed significant pulmonary venous desaturation.

Of our cohort, 2 patients died prior to being listed for transplant. The remaining 5 patients have undergone successful transplants. They all had resolution of their hypoxaemia before discharge from hospital post-transplant and have remained without supplemental oxygen. Their mean follow-up time is 73 months.

We studied 1 female, aged 13 years, with a diagnosis of PoPH. She presented with exertional dyspnoea and syncopal episodes. She had cyanosis on exertion with SpO_2 of 96% at rest. The work up for pulmonary hypertension incidentally revealed a portosystemic shunt (Portal vein to Inferior Vena Cava) on ultrasound imaging. Cardiac MRI demonstrated mean pulmonary artery pressure of 43mmHg at diagnosis which subsequently improved to 22mmHg with triple combination therapy (oral phosphodiesterase inhibitor, oral endothelin receptor antagonist and inhaled prostacyclin). Currently, she is being consider for portosystemic shunt closure.

Transplant remains the only cure for HPS and in this series hypoxaemia resolved in all patients. Patients with the lowest oxygen saturations at presentation had poorer outcomes (i.e death). Further research is required in PoPH to determine efficacious treatment strategies and ascertain the role of liver transplantation in this cohort.

HPS and PoPH are rare entities associated with very high levels of mortality if left untreated, hence the need for further research to aid early clinical diagnosis and curative treatment strategies for these patients.

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OC52

Biliary Atresia family focused pathway is essential in supporting families through the patient journey

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Evidence from literature suggests a huge psycho-social impact (emotional, financial, relationships, work, leisure) on parents and families of having a child with a health condition, with family response influencing both the course of illness and family's health and happiness. There can be positive aspects such as family relationships becoming stronger and post traumatic growth.

A decision was made to offer a family centered approach (systemic psychotherapy) specifically to all newly diagnosed BA families with focus on parental wellbeing, coping, impact on relationships and strategies to help them moving forward.

The pathway began at the end of 2022 / beginning of 2023 and has run until October 2024. There were patients not offered this intervention for agreed reasons (e.g. already engaged in HiM input, palliative involvement) and there were a small number who were unintentionally missed. During this time 25 families were offered input, and 21 accepted support. Of this, 18 were met F2F at diagnosis and 3 were not met at diagnosis but followed up shortly after.

Of the 21, ongoing support (defined as more than just meeting at diagnosis) was participated in by Mum only (7) and both parents (10). Families were seen as inpatients and outpatients and sessions completed virtually and F2F, this was agreed collaboratively with parents and flexible to their changing needs.

The common findings with specific themes around patient journey are as follows : -

- *A lack of research in this area.*
- *The role of fathers – they are likely to be struggling emotionally but in different ways and this may get missed.*
- *Impact on relationships – there are additional burdens and tensons and a 'medicalizing' of parents.*
- *Changes in individuals – self-identity, career, day to day life.*
- *Making meaning and 'ambiguous loss'–adjusting expectations and exploring new /alternate possibilities. Redefinition of their relationship with the situation and explore parental perceptions about the illness.*

Themes

At diagnosis / Kasai:

- Parental guilt.
- Anger of not being listened to pre BWCH.
- Adjusting to being in hospital, working with the team, being away from support network.
- Adjusting to being a parent of a child with a health condition.
- Grief and loss of hopes and expectations.
- In 'survival' / 'threat' mode.
-

Post Kasai / after discharge:

- *Living with uncertainty.*
- *Renegotiation of roles – as a partner, parent of an unwell child.*
- *Reorganisation of day to day and longer term.*
- *A need for self-care / external support.*
- *Struggle to acknowledge their own needs.*
-

Longer term:

- *Continuing to live with and adjust to uncertainty.*
- *Preparing for transplant / undergoing a transplant/*
- *Role – parent / carer / medic / organiser.*
- *Reorganisation of day-to-day and longer-term life.*
- *Sustaining themselves and each other.*
- *Balancing the urge to protect child with their growing independence.*

There is potential for preventative work with parents, resilience development and creation of resources to support parents with containment early in their journey, but also to sustain in long term with family therapy group pathway.

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OC53

Safety and Efficacy of Carvedilol in Children with Portal Hypertension: A Single-Centre Retrospective Study

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Carvedilol, a non-selective beta-blocker widely used in adult portal hypertension (PHN) [1], has limited evidence for its efficacy in paediatrics.This retrospective cross-sectional study from a paediatric liver centre in the United Kingdom evaluated Carvedilol's tolerability and efficacy in managing paediatric PHN.All children who commenced Carvedilol for PHN were identified from the Liver unit data base. Those with minimum follow up of one year post Carvedilol were included. Post Transplant and three other patients were excluded: two due to adverse events (dizziness and hallucinations in the first week of commencing Carvedilol) and one for non-adherence.Demographics, laboratory data, diagnosis, Fibro scans, shunt surgery, frequency of endoscopy and need for intervention (banding and /or sclerotherapy) were obtained from medical records. All children were on endoscopic surveillance and had required intervention (banding and/or sclerotherapy)before starting Carvedilol.Screening before commencing Carvedilol, dose of Carvedilol and frequency of endoscopy was as per the unit protocol.The final cohort included 36 children on Carvedilol, (20 males, 16 females) with a median age of 10 years (range 1-16). 20 children had cirrhotic portal hypertension (CPHN) and 16 non-cirrhotic portal hypertension (NCPHN).The median duration of endoscopic surveillance before and after starting Carvedilol was 4.38 (0.28-9.26) and 1.1(0.38-3.32) years in the NCPHN group, and 2.31(0.24-9.58) years and 1.5(0.23-2.7) years in the CPHN group respectively.Fibro scan readings were available in 16 patients (8 in each group) before starting Carvedilol, with median values of 20 (7.4-48.2) kPa in CPHN and 6.16 (2.7-10.2) kPa in NCPHN groups.Breakthrough bleeding was noted in 3 out of 16 patients in the NCPHN group compared to 2 of 20 patients in the CPHN group post Carvedilol. A significant reduction in endoscopic interventions in the CPHN group (p=0.011) following carvedilol therapy was noted. Of the patients with break through bleeding, 1 patient in the NCPHN group required liver transplantation and 2 had shunt surgery.Both patients with break through bleeding in the CPHN group had unsuccessful Trans jugular intrahepatic portosystemic shunt placement.One remains well on Carvedilol and the other had embolization of gastric varix.The study has several limitations: missing data points, small sample size and a short follow-up after Carvedilol initiation. Despite these limitations, the findings demonstrated good medication tolerability and promising results in reducing endoscopic interventions in the CPHN group.The authors suggest longer follow-up studies to establish Carvedilol's efficacy in paediatric PHN management.

Table 1: Effect of Carvedilol in study population.

	Mean+/- SD		P value	95% Confidence Interval
	Before commencing Carvedilol	After commencing Carvedilol		
Non cirrhotic portal hypertension n= 16				
APRI	1.1+/- 0.6	1.5+/-1	0.121	[-0.1325, 1.0263]
Fib-4	0.7+/-0.4	1+/-0.50	0.173	[-0.1081, 0.5494]
Endoscopies per year	3.4+/-3.3	3.1 +/-1.25	0.761	[-4.0735, -1.505]
Endoscopic intervention per year	2.8+/-3.4	1.5 +/-1.4	0.158	[-3.1763, 0.5662]
Cirrhotic Portal Hypertension n= 20				
APRI	2.5 +/-2.8	3.6+/-2.6	0.033	[0.08683, 1.9762]
Fib-4	1.4+/-1.2	2.2+/-1.8	0.018	[0.1484, 1.4526]
Endoscopies per year	3.6+/-3.3	2.2+/- 1.6	0 .08	[-2.0845, 1.5552]
Endoscopic interventions per year	2.6 +/-3.4	0.7+/-0.7	**0.011**	[-3.2066, -0.4837]

APRI: Aspartate aminotransferase to Platelet ratio, Fib-4: Fibrosis -4, SD: Standard deviation

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OC54

Novel Technique for Management of Post-Transplant Refractory Diarrhoea in Children with PFIC type 1

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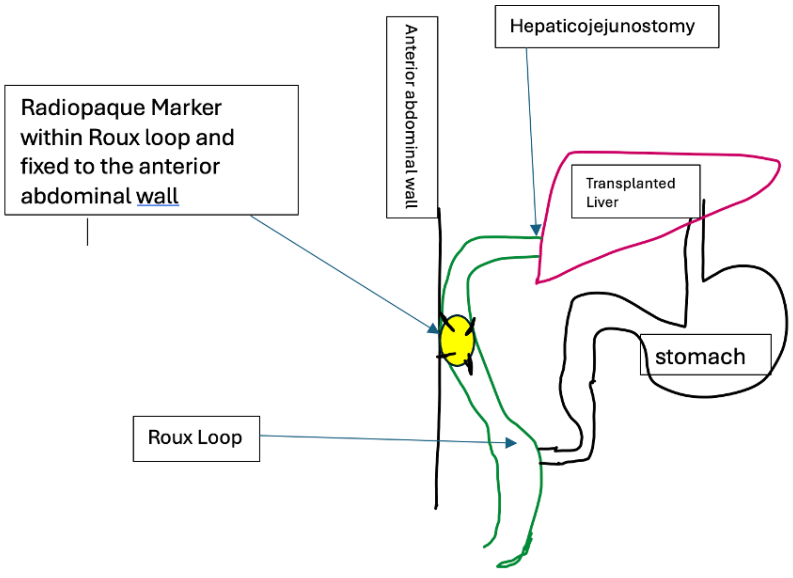
Progressive familial intrahepatic cholestasis type 1 (PFIC1), or Byler disease, is a rare genetic disorder that disrupts bile flow in children [1].Although liver transplantation (LT) offers a curative option, post-LT patients with PFIC1 often experience complications like chronic refractory diarrhoea and graft steatosis due to bile acid malabsorption [2,3].Surgical biliary diversion is commonly used before or after LT to alleviate symptoms such as severe diarrhoea and pruritus by reducing bile acid load [4]. Recently, ileal bile acid transporter (IBAT) inhibitors have shown promise as a non-surgical alternative for managing symptoms in PFIC1 patients without prior LT [5].However, not all PFIC1 patients require biliary diversion post-LT, due to variability in symptom presentation and timing of complications in addition to the unclear full pathophysiology of the disease. To address these challenges, we developed a novel technique for marking the Roux loop during transplantation.This marking allows for minimally invasive access to the Roux loop for surgical biliary diversion if it becomes necessary post-transplant, providing a flexible and less invasive management option for these patients if needed.This retrospective observational case series involved three paediatric patients with PFIC1 who underwent liver transplantation at ages 2.5 years, 3 years and 7 months, and 4.5 years respectively .All patients underwent transplantation due to end-stage liver disease. Two of these patients had biliary diversion pre transplant(one required it post transplant as well), and 1 case did not have biliary diversion before or after transplant .All three cases had a novel technique of marking their Roux loop by radiopaque marker during the LT procedure (Figure1).This marking aimed to facilitate less invasive future interventions if biliary diversion (BD) was required. Of the three patients, one developed severe refractory diarrhoea with severe hepatic steatosis post-LT and underwent partial external biliary diversion via interventional radiology, utilizing the pre-marked Roux loop without the need for major surgery. The remaining two patients did not need biliary diversion at the time of the study, as they exhibited only mild hepatic steatosis and their diarrhoea was not severe. In the case requiring biliary diversion (performed 7 months after second liver transplantation due to severe watery diarrhoea), the patient exhibited significant clinical and histopathological improvements following the procedure.The frequency of bowel movements decreased as well as the loperamide dose. Liver histology showed a reduction in hepatic steatosis from severe to moderate, with no progression of fibrosis. Due to stomal complications and patient preference, the external biliary diversion was later converted to an internal/external biliary diversion with a jejuno-colonic anastomosis.After this modification, the patient demonstrated good weight gain and reduced stool frequency to 2-3 times daily.Marking the Roux loop enabled efficient and minimally invasive access to the biliary system for the intervention, avoiding more invasive surgical approaches.**In conclusion,** marking the Roux loop during liver transplantation for PFIC1 is an innovative technique that allows for simpler, less invasive management of post-transplant complications. Further research is needed to confirm these benefits in larger patient groups.

Figure1 .Radiopaque marker in Roux loop for post-transplant access



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OC55

Evaluating alpha fetoprotein as prognostic marker for liver transplantation in paediatric liver failure

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In paediatric acute liver failure (PALF), risk prediction models are unable to predict survival with native liver with certainty. Alpha fetoprotein (AFP) has been proposed a promising prognostic biomarker and has been evaluated in neonatal and adult ALF but has not been assessed in PALF[1,2].

We aimed to evaluate AFP as a prognostic marker for survival with native liver (SNL) in PALF (prothrombin time >20 not corrected with vitamin K and liver dysfunction) and compare this to the previously validated CHLA-acute liver failure (CHALF)[3] which uses biochemical values to predict SNL versus need for Liver Transplant (LT) or mortality.

This was a UK health research authority approved single centre retrospective cohort study. Patients were identified from the departmental PALF database from Jan 2022 to August 2024 and included if they had at least one AFP measurement during their admission. Initial AFP, maximal AFP and initial:maximal AFP ratios were recorded. CHALF score was calculated on admission or when sufficient biochemical values were available.

AFP was measured in 19 of the 37 patients transferred to tertiary liver unit with PALF. Median (IQR) age was 3.5 (2.4-4.2) years and 9 were female. 6 children had a single AFP measurement and others had up to 10. 11 patients required LT and 1 died.

Median first AFP was similar (SNL: 53(26-129), non-SNL: 50(16-155); p=0.87). Maximum AFP and median AFP ratio were higher in the SNL group (medians 583 vs 111 and 7.70 vs 3.97) but this was not statistically significant. Median CHALF score was significantly lower in the SNL group (35 vs 38, p = 0.02). First AFP, maximum AFP and AFP ratio performed poorly compared to the CHALF score (area under receiver operating characteristic curves (AUC) 0.53, 0.73 and 0.62 vs 0.81). Using univariate logistic regression, a coefficient for first AFP was calculated and combined into the CHALF score calculation yielding an AUC of 0.84.

This analysis shows that the CHALF score performed better than AFP measurements when used in isolation. The inclusion of AFP into the existing CHALF score may improve its performance further. While maximal AFP and AFP ratio were higher in the SNL group, the differences were not statistically significant. However this cohort was too small to draw meaningful conclusions and data should be considered hypothesis generating only. Dynamic AFP trends have been demonstrated to be prognostic in adults and neonates. Incomplete and variability of repeated AFP measurements in this dataset were important limitations. Caution needs to be exercised as many laboratories in the UK use the wet enzymatic method for ammonia estimation which may give erroneously high levels in presence of high transaminases[4]. The external quality assessment (EQA) programs have attempted to harmonize AFP estimation[5]. Further work should concentrate on evaluating AFP in a larger PALF cohort, measuring repeat AFP at defined time points and assessing if AFP may be a useful addition to existing risk prediction models.

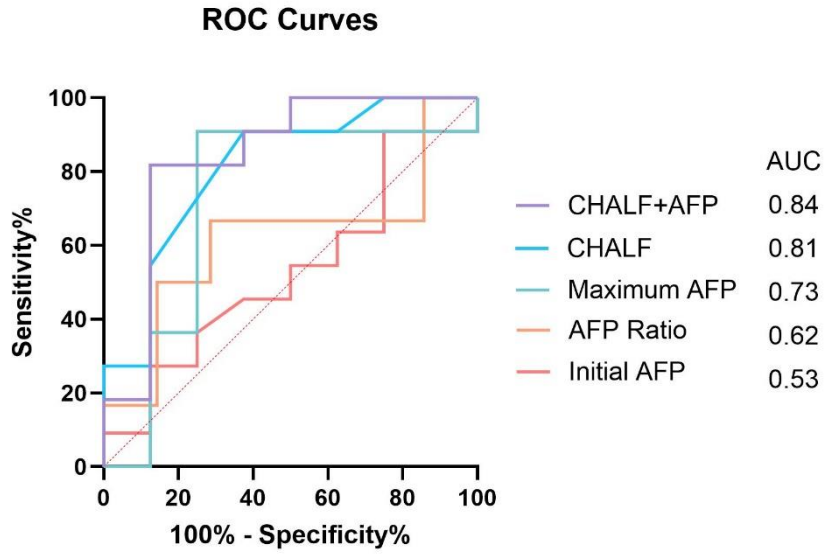

Figure 1 - Receiver operating characteristic curves for initial AFP, AFP ratio, maximum AFP, CHALF and CHALF + AFP with area under the curve indicated.

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OC56

Anaemia and red blood cell transfusion in paediatric liver transplantation: A retrospective observational study

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Liver transplantation (LT) is the standard treatment for children with end-stage chronic liver disease (CLD). Packed red blood cell (PRBC) transfusions are often needed to manage anemia pre- and post-LT, as well as blood loss during LT surgery. However, transfusions can increase sensitization risks, leading to adverse outcomes[1,2]. This retrospective study aimed to assess the incidence of PRBC transfusions in children with CLD during the perioperative period at a UK LT center. Since anemia is common in these children[3], a secondary aim was to investigate if they were screened for nutritional anemia and treated for deficiencies before transplantation.

The study included children with CLD who underwent LT between June 2021 and February 2023. One child was excluded due to care being transferred to another center. Children listed for acute liver failure or hepatoblastoma were excluded.

Data was collected at three points: perioperative (from listing for LT to surgery), intraoperative (during surgery), and postoperative (up to 28 days post-LT). Screening for nutritional anemia included iron studies, vitamin B12, and folic acid levels, using laboratory age-specific reference ranges for hemoglobin and mean corpuscular volume (MCV). Data was sourced from medical and blood bank records.

Among 29 children analyzed, the median wait time for LT was 49.5 days (range: 1–254 days), with 48% (n=14) being female. Biliary atresia was the most common underlying cause (48%, n=14).

Of the patients, 27 were anemic. PRBC transfusions were needed in 10% (n=3) preoperatively, 89% (n=26) intraoperatively, and 58% (n=17) postoperatively. None of the transfusions in the preoperative or postoperative periods were due to bleeding.

In the preoperative period, one of the three children requiring PRBC transfusions was screened for iron deficiency anemia (IDA) and found deficient; however, no iron supplements were given. None of these patients were screened for vitamin B12 or folic acid deficiency.

Table 1 shows the results of nutritional anemia screening and supplementation if deficient. Ferritin levels were tested in 10 children, with 9 showing normal levels. One child with low ferritin had IDA based on iron studies but normal MCV.

This study reports the incidence of PRBC transfusions and inadequate screening for nutritional anemia in children with CLD undergoing LT. MCV and ferritin levels alone are insufficient for diagnosing nutritional anemia. Iron studies, along with vitamin B12 and folate assessments, may help differentiate anemia of chronic disease and nutritional anaemia and may help reduce PRBC transfusions preoperatively.

Table 1: Screening and treatment of children with anemia while awaiting Liver Transplant

n=27	Screened with iron studies	IDA/treated	Screened for Vitamin B12 deficiency	Vitamin B12 deficient/ treated	Screened for Folate deficiency	Folate deficient/ treated
Low Hb & low MCV (n=4)	2	2/1	2	0/0	2	1/0
Low Hb & normal MCV (n=20)	13	3/2	7	0/0	11	6/3
Low Hb & High MCV (n=3)	0	0/0	1	0/0	1	0/0

Hb: Hemoglobin, MCV: Mean Corpuscular Volume, IDA: Iron deficient anemia

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OC57

Paediatric liver transplantation in tight junction protein 2 (TJP 2) deficiency

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Progressive familial intrahepatic cholestasis (PFIC) is a heterogenous group of inherited autosomal recessive disorders which is known to cause chronic cholestatic liver disease, often requiring liver transplant (LT)[1]. Homozygous and bi-allelic compound heterozygous mutations TJP2 protein results in TJP2 deficiency which is also known as PFIC4[2]. TJP2 deficiency causes disruption of tight junctions leading to progressive chronic liver disease. Extrahepatic manifestations of TJP2 deficiency includes subdural haematomas and chronic respiratory disease. There is little evidence on the outcomes of liver transplantation in TJP2 deficiency.

The objective of this study was to describe the clinicopathological features and posttransplant course of six LT recipients with TJP2 genetic mutation (PFIC 4). Data was retrospectively collected from electronic case records of children (1-18 years) with TJP2 deficiency, who underwent LT between March 2008 and December 2013.

We identified 6 patients with genetically confirmed TJP2 deficiency. 4/6 patients presented with neonatal jaundice. One patient presented at 5 years of age with focal seizures and deranged Liver function tests. One patient presented with pruritus. The median (IQR) bilirubin on presentation was 108.5 (83-131.2). 3/6 patients had severe pruritus for which one patient required external biliary diversion. 4/6 patients had undergone liver transplant and the remaining 2 patients had milder phenotype and have been transitioned to adult services. All the transplanted patients were homozygous for pathogenic mutations. The median age at transplantation was 11.5 years. The outcomes of LT are listed in table 1. One patient was transferred to another centre for LT and hence no long-term data was available. All three patients under our care recovered uneventfully after LT. One patient had to undergo a re-transplant in 2017 due to recurrent cholangitis and unfortunately died of multi-organ dysfunction due to sepsis at 19 years of age.

Table1: Pre-transplant profile and post liver-transplant course

Variable	Patient 1	Patient 2	Patient 3
Sex	F	M	F
Genetic mutation	TJP2: c.1099C>T; p.(Arg367Ter)	homozygous c.813delC	homozygous c.813delC
Type of mutation	Stop gain change	pathogenic frameshift	pathogenic frameshift
Pruritus	Yes	Yes	Yes
Explant histopathology	Cholestatic Giant Cell Hepatitis	Cirrhosis	Cirrhosis Early Hepatocellular carcinoma
Age at LT (years)	1	10	14
Indication for LT	End stage liver disease	End stage liver disease	End stage liver disease
Follow up in months post LT	120	98	56
Post operative complications	Hypertension, Chylous ascites	Small bowel obstruction due to adhesions, Recurrent cholangitis	Splenic artery embolization for steal phenomenon, Hepatic vein anastomotic stricture
Graft Rejection	Yes	Yes	Yes
Retransplant	No	Yes	No

TJP2 deficiency is a rare cause of molecular cholestasis which can result in progressive endstage liver disease needing liver transplant. The severity of liver disease correlates with the type of change caused by the mutation. No functional protein leads to very severe disease requiring LT early in their life. LT experience in TJP2 deficiency is not widely reported and this case series adds to the evidence.

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OC58

Transition services in paediatric inflammatory bowel disease: a single tertiary centre experience

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Transition from paediatric to adult care is a critical phase for adolescents with inflammatory bowel disease (IBD). 20-30% of patients are diagnosed with IBD before the age of 20, and paediatric-onset illness has shown to be associated with greater disease severity[1,2,3]. It is essential to optimise management prior to transfer to adult services. Transition services provide pivotal opportunities to empower patients with greater responsibility and autonomy within a well-supported environment. There is limited literature regarding the transition process, particularly from a patient-centred view, and a lack of standardised approach to transition services[4,5,6].

This study aimed to evaluate the current IBD transition service at a single tertiary centre and identify any recurring themes and targets for improvement within service delivery.

50 patients aged 16-18 were selected retrospectively from the departmental IBD database. Electronic patient notes were evaluated and data collected on age, sex, diagnosis and treatment. Thematic analysis using open coding was performed on consult notes and letters to identify transition-related themes.

Mean age was 16.5 years, 26/50 (52%) were male and 24/50 (48%) female. 35/50 (70%) had Crohn's disease, 10/50 (20%) had IBD-unclassified and 5/50 (10%) had ulcerative colitis. 34/50 (68%) were on thiopurines, 31/50 (62%) were on aminosalicylates and 19/50 (38%) were on biologics. 2/50 (4%) were on no maintenance therapy. 32/50 (64%) were in clinical remission.

31/50 (60%) had a completed transition clinic appointment. In 4/31 (13%) the parent was present without the patient. 6/50 (12%) had failed to attend at least 1 transition appointment. 4/50 (8%) explicitly expressed they did not feel ready for transition, most commonly due to active disease.

Mental health issues affected 12/50 (24%); 5/50 (10%) reported anxiety, 3/50 (6%) reported eating disorder/poor self-image behaviours and 4/50 (8%) reported autism as having a significant effect on their well-being and disease management. 9/50 (18%) reported school absences having a substantial effect upon their work and examinations. Future education or work was discussed in 36% of consultations.

When prompted, 8/13 (62%) had medication compliance issues with reasons including poor understanding and forgetfulness quoted.

Menstrual cycles were broached in 25% of female consultations, with 80% of these patients reporting irregular periods. 1/50 recorded discussion of smoking. Sex, relationships, alcohol and substance use were not discussed.

20/50 (40%) wanted to continue care within the Trust's adult services, 10/50 (20%) at a local district general hospital (DGH) oand 20/50 (40%) had not indicated a preference yet.

Key issues at the time of transition to adult IBD services include mental health, school and education, and medication. Particular consideration should be given to these in clinic. Prevalence of mental health issues is comparable to reported rates of 22% in the general adolescent population[7]. Relationships, sex, substance use, alcohol and smoking were rarely discussed. Incorporating a structured tool into clinic review such as the HEADSSS assessment[8] can help clinicians structure discussion of these and mental health issues. Barriers to patient attendance at clinic and patients' views on service design should be sought to ensure young people attend key clinic appointments.

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OC59

Is adrenal insufficiency a risk in children with inflammatory bowel disease treated with systemic steroids? Our experience interpreted through NICE guideline.

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Adrenal insufficiency (AI) is a recognized complication following systemic steroid therapy in paediatric patients, including inflammatory bowel disease (IBD)[1,5]. Nevertheless, review of guidelines from relevant societies (BSPGHAN, ESPGHAN, and ECCO) shows no formal, standardised management pathway for monitoring AI in these patients. This study aims to identify possible risk factors for AI in children with IBD receiving systemic (intravenous or oral) steroid therapy, and to propose a screening and management protocol. Between June 2022 and June 2024, this single-centre, prospective study evaluated 9 children with IBD receiving more than two courses of systemic steroids out of 100 patients cared for within this time frame. Eight patients were investigated, six of whom had a low early morning free cortisol level suggestive of AI. No correlation was observed between insufficient early morning cortisol level and type of IBD, clinical characteristics at diagnosis, type or dosing of the systemic steroid treatment received (Table 1). Within this group, symptoms were nonspecific or absent, emphasizing the difficulty of predicting AI without screening. Children in this group were treated with supplemental oral hydrocortisone and were followed up 3 monthly. By the end of the observation period , 1/6 children had regained a sufficient cortisol level after 8 months. In line with the most recent NICE guidelines highlighting a risk for AI in children receiving more than 3 weeks of systemic steroids, [4] we have identified a relevant prevalence in this study population similar to previous studies[1,2,3] and therefore support recommendation for AI screening via early morning cortisol level in this context. We recognise the limitation of small numbers in this study and aim for a multi-centric collaboration on a national level to develop a comprehensive pathway based on the one proposed here.

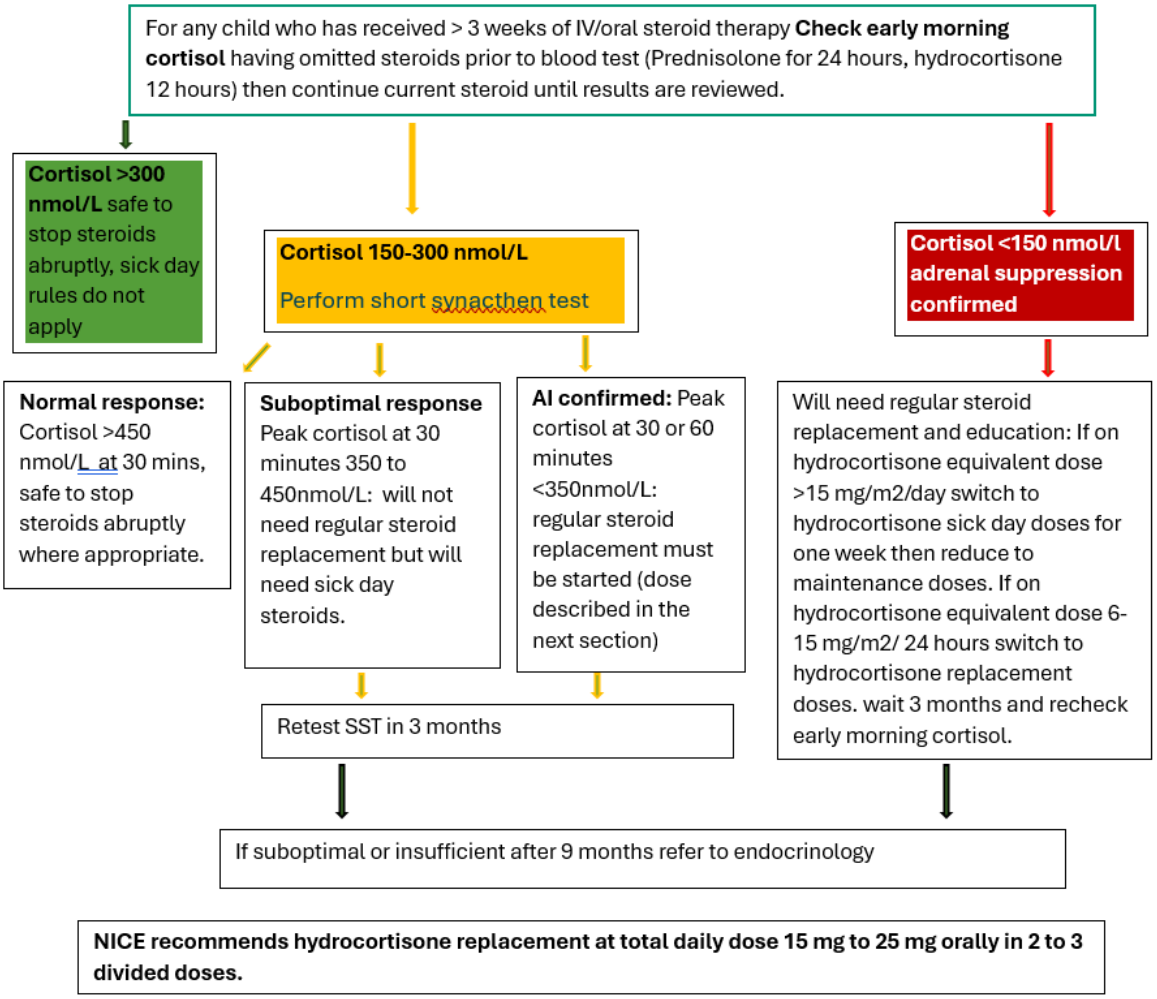
Table 1 Patient data from our study.

Patient Status	**Number of Patients**	**Steroid Type**	**Starting steroid Dosage (mg/m²)**	**Courses of Steroids (more than 3 weeks)**	**Symptoms**	**Type of IBD**	**Age at diagnosis of IBD** **A1a: 0 to < 10 yrs** **A1b: 10 to < 17 yrs**
Adrenal Insufficient	6	IV hydrocortisone/oral prednisolone	20-48	2-8	Tiredness, body aches, no symptoms.	CD n=2 IBD-U n=2 UC n=2	A1a n=4 A1b n=2
Adrenal Sufficient	2	IV hydrocortisone/oral prednisolone	25-30.7	2-3	Weakness, dizziness, low energy, or no symptoms	CD n=1 UC n=1	A1a n=0 A1b n=2

Table 1 Legend: CD: Crohn's disease; IBD-U: inflammatory bowel disease unclassified; IV: intravenous; UC: ulcerative colitis.

Suggested pathway for investigating and managing AI post steroid therapy

OC59 - continued



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OC60

Postal faecal calprotectin testing: a qualitative study assessing impact of a pilot on patient and carer experience in a Paediatric Inflammatory Bowel Disease (PIBD) population.

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There are several challenges that come with the care of adolescent patients when managing long term conditions, including Inflammatory bowel disease (IBD), which can have a negative impact upon adolescent patients' sense of self [1]. Management and monitoring of IBD shows tighter control with regular faecal calprotectin monitoring [2]. Even in asymptomatic patients, it is a proven way to monitor disease activity and predict symptomatic relapse [3]. It is recognised, that there is an issue with non-adherence and that patients find it challenging to autonomously provide regular samples [4].

Within an urban, tertiary paediatric gastroenterology unit, several patient barriers to stool sampling were identified from informal family feedback. These included the reluctance for in-hospital sample collection; embarrassment of walking around carrying samples; inconvenience and time spent away from family/work for carers in physically dropping off samples; samples getting lost after drop-off.

With the aim of improving the sampling process for patients, a postal sample service has been established. Postal samples are already used successfully nationally in the Bowel Cancer Screening Programme [5,6].

Families are given packs containing sample kits which can be posted back in a regular post-box direct to the laboratory.

To evaluate patient and carer acceptability of the intervention, qualitive data will be collected in the form of surveys. Data will be gathered on how families find the process; the ease of sample collection, whether samples have been lost and whether carers needed to re-arrange work or childcare to facilitate sample drop off.

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OC61

Rectolabial fistula as first presentation of Crohn's Disease in an adolescent

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Crohn's Disease (CD) is a chronic condition characterised by transmural gastrointestinal inflammation from oropharynx to anus.[1] Extra-intestinal manifestations of CD can occur in up to 40% of patients, spanning musculoskeletal, mucocutaneous, and ophthalmic involvement.

Whilst perianal and recto-vaginal associations of disease have been well-described in the literature,[2] rectolabial fistula is a rare extra-intestinal manifestation of CD, described only once prior in the literature, in the case of a 9-year-old pre-menarchal female.[3]

This report presents the unique diagnostic and management challenges in a previously fit and well 14-year-old Caucasian female with rectolabial fistula as first presentation of CD.
The patient presented acutely with labial pain and swelling. She reported 2kg weight loss and menorrhagia in the few months prior to presentation. This was on a background of chronic crampy abdominal pain, and loose bowel motions twice daily with blood on wiping and mixed with stool. She denied rectal mucus or other features of systemic involvement including fevers, arthralgia, eye, or skin involvement. She was not sexually active at any time prior to presentation.
Initial blood tests including ESR, CRP and FBC were within normal limits. Faecal calprotectin was 447 ug/g.

The patient was initially admitted under the gynaecology team for possible labial cyst. She underwent examination under anaesthesia (EUA), which excluded labial cysts but showed possible perianal fistula.

She was transferred to a tertiary centre for further assessment and management.

MRI perineum showed an area of oedema in the anterior right labia. A thin fistula at 11 o'clock was noted to pass posteriorly towards the anus (*Figure 1*).

Considering these findings, a multi-disciplinary investigative approach was undertaken incorporating GI endoscopies and rectal EUA.

Endoscopic appearances were in keeping macroscopically with CD. Colonic biopsies confirmed moderate to mild active chronic colitis with granulomas.

EUA rectum confirmed anterior trans-sphincteric fistula between right labium and rectum, involving 50% of internal sphincter without evidence of recto-vaginal fistula, abscess or collection. An anterior seton was inserted.

Labial biopsy showed multiple dermal non-caseating granulomata associated with a predominantly chronic inflammatory cell infiltrate. No dysplasia or malignancy.

After initial treatment with antibiotics, the patient commenced infliximab 4 weeks post-seton insertion. The patient completed a 3-month course of oral Metronidazole.

Repeat EUA 6-months following the first confirmed persistence of the fistula involving 40% of the sphincter, with development of granulation tissue. Partial fistulectomy and seton change were performed.

As well as evidence of clinical remission, MRI perineum at 1 year showed complete fistula resolution.

Alongside clinical complexity, this patient suffered with the psychological burden of this chronic disease; she had significant school absence that correlated with the fluctuating severity of physical symptoms and psychological distress. She was offered psychological and educational support.

Key messages
A presentation of labial swelling and tenderness may warrant further work-up for CD in a paediatric population.

Effective clinical management requires close multi-specialty collaboration including paediatrics, gastroenterology, gynaecology, radiology and general surgery.

The biopsychosocial burden of this chronic disease with gynaecological manifestations presents a unique challenge, mandating multi-modal support including psychologists, school and family.

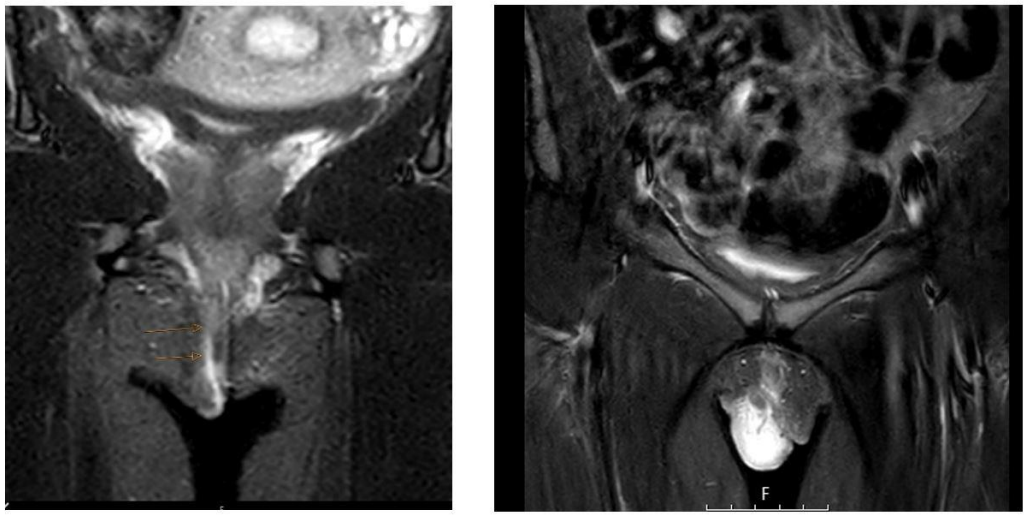

Figure 1: MRI perineum showing fistulous tract and labial swelling, arrows in left image indicate the fistula

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OC62

Upadacitnib in the treatment of paediatric refractory inflammatory bowel disease; experience at a specialised paediatric UK centre

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Upadacitinib (UPA) is a novel orally administered small molecule drug that works as a selective Janus kinase (JAK) inhibitor and is approved for use in adults with ulcerative colitis (UC) and Crohn's disease (CD)[1]. There is increasing off-licence use of UPA in paediatric patients but there remains little data of its safety and efficacy[2-4].

We present here early data on use of UPA over 12 months in an adolescent cohort with refractory IBD at a single centre from October 2023 - 2024. We identified patients ≤ 18 years with IBD started on UPA and gathered data on demographics, disease subtype, number of biologic failures, steroid use, adverse events and biomarkers of remission.

17 patients with paediatric IBD (median age 16 years, range 12-17 years old, 9 females (53%)) were identified. 4 patients had UC and 13 had CD. 8/17 (47%) commenced UPA following failure of 2 biologics, 6/ 17 (35%) had failed 3 or more biologics (1 had trialled 4 drugs: adalimumab, infliximab, ustekinumab, vedolizumab). In all patients previous biologics were stopped and UPA was used as monotherapy. All patients had active disease on commencement of UPA; 3 out of 17 were on steroid treatment and all had faecal calprotectin >200µg/g. 5/17 patients (29%) had a high C-reactive Protein (CRP), only 1/17 patients had a low albumin. All patients were started on the recommended 45mg UPA.

By patient and physician global assessment 13/17 patients (82%) had clinical improvement/clinical remission at 4-8 weeks of UPA therapy. For 3 patients on steroids, weaning was successful and no further courses required. Of 13 patients whose results were available at time of data collection, endoscopic remission, suggested by faecal calprotectin <100µg/g, was achieved in 7 out of 13 patients (53%) at 4 weeks of UPA therapy. One patient discontinued UPA at 3 months after induction therapy due to primary non-response. Relapse at 12 weeks following induction therapy was noted in 1 patient after dose decreased to 15mg (parental choice), then achieved remission on 30mg maintenance dose. All other patients were prescribed the higher maintenance dose of 30mg.

No serious adverse events occurred; 1 patient with a background of asthma stopped and restarted UPA at a lower dose following increased respiratory sputum burden and emergency department attendance, UPA was subsequently discontinued at 6 months for similar symptoms. 1 patient experienced a pruritic rash on first administration which resolved and did not recur, and 2 patients experienced acne but continued the medication.

In summary, this study showed most patients on UPA for refractory IBD had good compliance, tolerance and symptom improvement. Currently, 4.8% of the total paediatric IBD cohort in the study centre is being treated with UPA; its increasing use is in part due to its oral administration and it avoids the use of bridging steroids. It would be beneficial to repeat this audit with increased patient numbers and additional data. In addition, with the expansion of biologic therapies available, there is a mounting case for a nationally held prospective database for paediatric IBD patients.

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OC63

Does checking thioguanine levels add to the treatment of paediatric IBD patients?

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Background

Azathioprine is widely used in the maintenance treatment of inflammatory bowel disease (IBD) in both children and adults. The use of thiopurines is linked to dose-dependent adverse drug reactions, which frequently result in dose adjustments or discontinuation of the medication (1). Despite the increasing use of thiopurines in paediatric IBD management and the potential benefits of monitoring 6-TGN and 6-MMP metabolites for assessing compliance and optimizing therapy, routine monitoring of these metabolites is not standard practice (2).

Methods

Paediatric patients with IBD who have been started on Azathioprine between 01.04.23 and 31.10.24 are enrolled in the study. Diagnosis date, dose and start date of Azathioprine were recorded. Thiopurine methyl transferase levels **(**TMPT) levels were checked. The levels of azathioprine metabolites, Thioguanine and MMP levels were checked approximately in 3 months.

Results

In total, 42 patients were enrolled in the study. Mean age of the patients at the diagnosis were 12.81±2.97 years old (range 4-16) 42% (18) were female. Out of 42 patients, 64.3% were Crohn's, 28.6% were Ulcerative Colitis and 7.1% were IBD-U. TPMT levels of 35 out of 42 patients were available. 4 (11.4%) of them had low levels of TPMT, 26 (74.3%) were in normal range (26-50) and 5 (14.3%) had high levels.

The thioguanine levels were available in 27 out of 42 patients. The average time for checking 6-TGN were 88.68±45.3 days (min:36 max 248). In 33.3% (9) of the patients, the levels were below the normal range, %26.9 (8) were above the range and 37% (10) were within range.

Conclusion

Azathioprine metabolite monitoring is more common in adult IBD management, as specific metabolite ranges are linked to improved response and reduced risk of adverse effects. (3,4). In paediatrics, it is not an established practice, however there are publications implying the usefulness and importance of metabolite monitoring in IBD maintenance treatment (5,6). In our cohort, the thioguanine levels in 33.3% (n=9) of the patients were below the therapeutic range and 3 of them had dose increase based on these results and the rest of them were reviewed from compliance point of view. Also 26.8% of the patients were found to have TGN levels above the therapeutic range.

The monitoring of TGN metabolites can be useful to optimise the IBD treatment in paediatric patients, not only in terms of avoiding toxicity and optimising the dose but also would be effective to assess the compliance which is a common obstacle in the treatment of teenage patients.

Tables

Table -1 Basic characteristics

Age (years)	12.81±2.97
Crohn’s	64.3% (27)
UC	28.6% (12)
IBD-U	7.1% (3)
Average time to check TGN level (days)	88.68±45.30
Average Azathioprine dose (mg/kg)	1.62±0.37

Table 2 Classification of patients according to thioguanine level

Thioguanine level	**percentage**
Between range (235 - 450 pmol/8x10^8 RBC)	37.0
Below the range	33.3
Above the range	29.6

Table - 3 Classification of patients according to TPMT levels	
Enzyme level	**percentage**
Carrier (10-25)	11.4
Normal (26-50)	74.3
High (>50)	14.3

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OC64

Facilitating and evaluating the beads of life workshop for young people with inflammatory bowel disease: *I recognised all the things I have overcome and that I wasn't alone.*

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The Beads of Life is informed by narrative principles, using beads to represent different aspects of an individual's life. It was initially developed for children and young people with cancer as a way of making sense of their cancer journey through building strength and resilience. It has also been adapted for other paediatric populations, supporting young people to change their relationship with their health condition where a difficult narrative may otherwise be dominant.
Given that children with IBD also report difficulties navigating and managing symptoms, treatment and overall wellbeing, the current group was adapted to support patients with IBD to share their preferred narrative, explore their IBD journey and connect with peers.

The Paediatric Gastroenterology Psychology Service ran four one-day group workshops from October 2023-October 2024 with 23 participants in total aged 10-17 with a diagnosis of IBD. When possible, the workshops were also co-facilitated by a young person who had previously attended. The workshop invited attendees to select beads that represent their preferred narrative including strengths, important people and hopes and wishes for the future. Using externalisation, attendees were able to understand their diagnosis from a different perspective which led to further beads selected to represent their IBD journey.

All attendees completed feedback on the workshop using a Likert rating scale alongside Qualitative feedback which was analysed using content analysis. Prior to the workshop and at 1 month follow up, attendees were asked to complete the Wellbeing and Health Experiences Evaluation Log (WHEEL) and IBD Self-Efficacy Scale for Adolescents and Young Adults (IBDSES-A).

On average, attendees rated sharing experiences of IBD as 8/10 (where 10 is 'Very Useful'), enjoying the group as 8/10 (Where 10 is 'Very Much Enjoyed'), and recommending the group as 8.3/10 (Where 10 is 'Very Much Would Recommend'). Themes from the qualitative feedback demonstrated the value young people placed on sharing their IBD experiences, relating and connecting to others as a way of reducing isolation and taking away messages of empowerment: *'Not having IBD hold me back.'*

The quantitative data of those that completed the WHEEL and IBDSES-A at the 1 month follow up was analysed using descriptive statistics (Mean; M, and Standard Deviation; SD). See table 1.

The feedback from the workshops is in line with previous findings using the beads of life approach. The themes support our aims to provide a safe space for children and young people to share their preferred stories of their identity and IBD journey, connect with others and reduce feelings of isolation: *'knowing you are not alone.'* This demonstrated promising results in supporting young people to make sense of their IBD journey.

Table 1

Mean scores for attendees that completed 1 month follow up questionnaires (*n*=8)

	WHEEL M (SD)	**IBDSES-A** M (SD)
Baseline	3.62 (0.35)	45.88 (3.80)
1 Month	3.89 (0.27)	48.38 (3.58)
Change	+0.27	+2.5

OC65

An initial review of upper oesophageal sphincter characteristics on high resolution oesophageal manometry

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While the lower oesophageal sphincter and oesophageal body have well defined normal ranges for manometric investigations in paediatrics, there is very limited data on the upper oesophageal sphincter (UOS) available for children, with most previous reports requiring significant additional manipulation of data outside of the commercially available software used to capture the data [1,2].

A retrospective review of oesophageal manometries carried out in a tertiary paediatric centre was carried out, with 30 patients randomly selected for additional upper oesophageal sphincter measurements to be extracted. Measurements of the upper oesophageal sphincter available within the Solar GI oesophageal manometry module (Laborie/MMS) were extracted, along with the baseline diagnosis following manometry. Achalasia has previously been shown to affect the upper oesophageal sphincter [3] so two patients in the random sample who were diagnosed with achalasia were excluded from the analysis.

28 patients were therefore included in the dataset, mean age 10.6±3.7 years, 11:17 M:F. The diagnostic outcomes of these patients were normal (12), rumination (4), ineffective peristalsis (8), belching (2) and reflux (2). The mean (±standard deviation) resting UOS pressure for 5ml swallows of rehydration salt solution was 44.8 ± 22.2 mmHg with mean residual pressure of 1.1 ± 5.5 mmHg. Mean IRP 0.2s was -0.7 ± 5.3 mmHg and mean IRP 0.8s was 10.6 ± 6.7 mmHg.

Ongoing work is underway to correlate manometric data with video fluoroscopy and other conditions as well as expand the sample to include all oesophageal manometries completed in the last 5 years to ensure access to a robust normal dataset for investigating children who present with symptoms of upper oesophageal sphincter dysfunction.

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OC66

Eosinophilic colitis in children with neuro-disability: A tertiary center's experience of four cases

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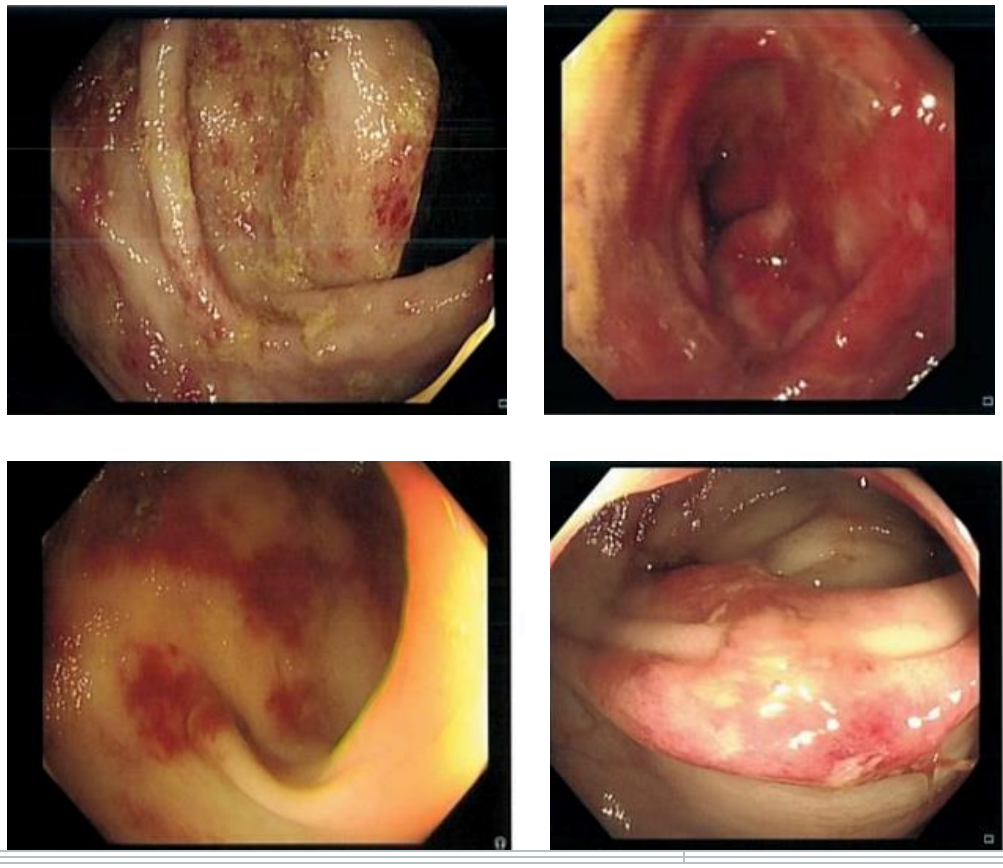
Eosinophilic colitis (EoC) is a subtype of eosinophilic gastrointestinal disorders (EGID) beyond eosinophilic esophagitis (EoE) which has been recently updated as a nomenclature (1). It describes chronic inflammatory disorders of the gastrointestinal (GI) tract characterized clinically by the GI symptoms and histologically by eosinophilic inflammation, in the absence of a secondary cause. EoC is the least understood of the non-EoE. EGIDs, as its clinical presentation can be confused with IBD (inflammatory bowel disease) such as ulcerative colitis or Crohn disease (2).

We report 4 patients who were referred to us with chronic per rectal bleeding, abdominal pain and diarrhea and raised faecal calprotectin. The median age was 11 years. They all had neuro-disability with severe development delay, non-verbal and were gastrostomy fed. Two of them had underlying syndromes. All of them were on hypoallergenic feeds with no allergic history. There were no drug associations found. On upper endoscopy and colonoscopy, there were patchy areas of submucosal hemorrhages which were mainly identified in the proximal colon. Macroscopically (Figure 1), they have not been described in EoC but correlated histologically with the suggested threshold peak eosinophil counts of EoC. One had associated EoE but the remaining did not have any upper GI changes. One of them was eventually diagnosed with Crohn's disease and subsequently passed away due to other, non GI related health issues. Rest were treated with sulfasalazine and are currently symptom free.

We concluded that faecal calprotectin can be elevated in children with EoC (3). The macroscopic findings at colonoscopy were unique to these children with neuro-disability. They were all on anti-epileptic medications but none of them are reported to be associated with EoC as an adverse effect. The presence of thresh-hold increased eosinophils in the colonic mucosa requires consideration for a secondary cause, especially IBD as EoC is rare and remains a diagnosis of exclusion. More data to ascertain if EGID is more common in neuro-disabled children is required since there is a lack of published data about EGID.

Figure 1: Macroscopic findings during colonoscopy showing submucosal haemorrhages



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OC67

The use of pylorus endoFLIP in children with symptoms of impaired gastric drainage

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The endoluminal functional luminal imaging probe (EndoFLIP) is a new and expanding area of medical technology. Currently its use is focused on investigating the function and motility of hollow organs of the gastrointestinal system, in the oesophagus, pylorus and rectum[1]. This study aims to retrospectively review the use of pylorus endoFLIP in children referred for symptoms of impaired gastric drainage in a single centre.

We retrospectively reviewed all patients (22) who underwent pylorus endoFLIP at a single centre between July 2022 and May 2024. Demographic data, indications, weight before and after, endoFLIP measurements, interventions received (botulin toxin injection into the pylorus) and symptoms after intervention were collected.

Of the included patients, there was an average age of 9 years and 5 months, and 32% were female. The percentage for each indication was: 90.91% vomiting, 68.18% retching, 13.64% abdominal distention, 54.55% intolerance to feeds and 22.73% pain after feeding.

11 patients received intrapylorus botox due to abnormal endoFLIP values. The group that received botox had an average age of 8 years and 7 months. The percentage of symptoms prior to and few months after botox injection were: vomiting (100% improved to 63.6%), retching (81.8% to 54.6%), abdominal distention (18.2% to 18.2%), intolerance to feeding (54.6% to 45.5%) and pain after feeding (27.3% to 36.4%). The average weight before and after was 22.88kg before and 23.30kg after intervention.

11 patients did not receive intrapylorus botox (due to normal endoFLIP measurements). They had an average age of 10 years and 4 months. The percentage indications were: 81.82% vomiting, 54.55% retching, 9.09% abdominal distention, 54.55% intolerance to feeding, 18.18% pain after feeding. Weight before was 26.82kg and after was 26.04kg. The table below displays the results of both intrapylorus botox and no botox, for diameter, compliance, pressure and distensibility.

Balloon Volume (ml)	Mean Diameter (mm)	SD Diameter	Mean Compliance (mm3/mmHg)	SD Compliance	Mean Pressure (mmHg)	SD Pressure	Mean Distensibility (mm2/mmHg)	SD Distensibility
20	9.02	2.31	151.98	97.22	13.57	7.81	5.33	4.48
	7.38	2.01	93.39	64.64	14.97	5.13	2.30	0.86
30	10.48	2.25	120.43	46.30	17.79	3.49	4.39	2.00
	8.72	1.91	82.07	37.12	24.28	13.57	2.82	1.41
40	14.23	2.77	131.08	59.99	35.44	7.21	4.90	2.39
	11.78	0.85	73.97	19.26	44.92	13.61	2.69	0.96
50	14.70	1.88	83.94	29.48	62.36	9.10	2.90	1.14
	13.22	1.38	197.95	211.97	53.38	28.92	3.70	2.51

White box = no intervention, yellow box = intrapylorus botox

Table 1: diameter, compliance, pressure and distensibility from endoFLIP in intrapylorus botox and no botox.

In conclusion, EndoFLIP is a promising tool in investigating pylorus dysfunction and symptoms of impaired gastric drainage. EndoFLIP can also be used to predict clinical response to intrapylorus botox injection.

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OC68

Assessment of Quality of Life Before and After Intervention in Patients with Achalasia - A Systematic Review

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Oesophageal achalasia is a rare gastrointestinal motility disorder of increasing incidence with no gender or racial predilection. It affects around 27.1 in 100,000 people in England and 10.82 in 100,000 globally and is rarer still in the paediatric population with an estimated incidence of 0.38/100,000 children per year.[1,2]

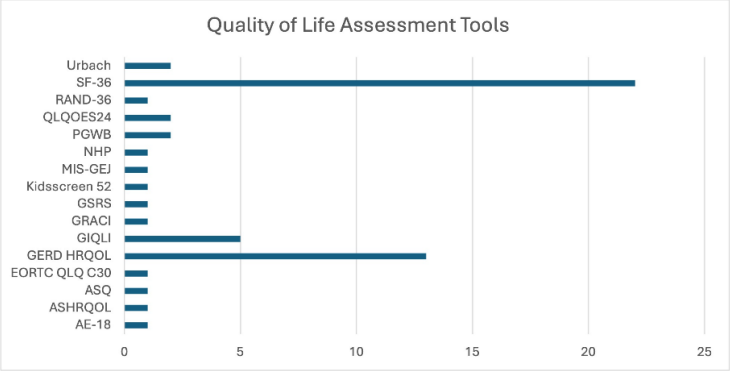
Achalasia causes ineffective oesophageal peristalsis and impaired lower oesophageal sphincter relaxation giving rise to the typical symptoms of regurgitation, dysphagia, retrosternal chest pain and weight loss.[3] In paediatric patients, food refusal and failure to thrive are commonly reported.4 The gold standard for diagnosis remains high resolution oesophageal manometry but diagnosis can be delayed due to the rare incidence combined with the overlap of symptoms and heterogenous clinical assessment tools.

Though relatively uncommon, achalasia is known to significantly impact health related quality of life (QOL). There is no curative treatment, but symptom amelioration can be achieved using interventions such as pneumatic balloon dilatation, laparoscopic Hellers myotomy and per-oral endoscopic myotomy.[4] Symptom improvement can positively impact QOL, however in patients for whom symptoms persist, mental wellbeing is poorer compared with the general population and may progressively worsen over time.[5]

Despite the recognition of the impact, there is no universally agreed disease-specific approach for evaluation of QOL of patients with achalasia. Additionally, evaluation of pre and post-intervention symptom profiles is inconsistent, further limiting the ability to understand the effectiveness of interventions.

We conducted a systematic literature review of existing research on quality of life in achalasia patients. Ovid was used to search multiple digital databases: Journals@Ovid Full Text Subset, Journals@Ovid, Ovid Medline (1946-2024), Embase (1974-2024), Ovid Emcare (1995 to 2024), HMIC Health Management Information Consortium (1979 to 2024).Initial literature search produced 1746 articles and following removal of duplicate articles and title screening 95 articles remained. Abstracts screening reduced this further to 69 and after full text review the resulting number of articles was forty-nine.

Predominantly these studies involved adult patient cohorts with only two articles (4%) evaluating children. The patient cohort sizes were variable with an average of fifty-four patients (range 8 – 201). Graph 1 demonstrates the significant heterogenicity of the QOL tools used, and despite the existence of disease-specific assessment tools (for example the Achalasia-Specific Quality-Of-Life-Questionnaire, ASQ) these do not appear to be widely implemented, suggesting either a lack of clinician awareness that these tools exist, or that the tools themselves are not fulfilling the clinical need.



Graph 1: QOL assessment tools

Considering the rarity of achalasia as a condition, this unstandardised approach significantly impacts the ability to draw meaningful conclusions about the impact of interventions on QOL. Furthermore, many studies did not use Ecktardt Score when evaluating the clinical symptoms, thus reiterating the lack of standardisation. Many do not include both pre and post intervention assessments, further adding to the deficit in understanding the clinical significance of intervention. To improve understanding of impact on QOL and thus outcomes for patients there is a need to develop a standardised QOL score to assess patients with achalasia before and after intervention.

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OC69

A rare case of achalasia in a 14 year old with oesophageal atresia and tracheoesophageal fistula

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We present a rare case of achalasia associated with oesophageal atresia (OA) and distal tracheoesophageal fistula (TOF). OA/TOF is a congenital abnormality due to incomplete embryonic development of the foregut. Achalasia is defined as insufficient lower oesophageal sphincter relaxation and absent peristalsis of the oesophagus, and has clear high resolution oesophageal manometry (HROM) diagnostic criteria. It has been postulated that there is an associated abnormality of the Auerbach plexus of the distal oesophagus and fundus of the stomach in patients with OA/TOF.

A retrospective case analysis and literature review of OA/TOF and achalasia was undertaken.

A 14-year-old girl was assessed at a tertiary paediatric surgical centre for vomiting. The patient had a complete OA with distal TOF repaired at 3 months of age. This was complicated by anastomotic leak, requiring further definitive surgery at 4 months. She also had complete heterotopic tracheal rings of the oesophagus which were excised at 14 months in addition to a partial fundoplication. She later presented with progressive dysphagia and was diagnosed with GORD.

She underwent time barium oesophagogram (TBE) at age 6 which demonstrated a long distal oesophageal narrowing, which was not evident at endoscopy. She had a total of 7 upper GI endoscopies which demonstrated distal oesophageal dilatation with gastric pooling and food debris but no stricture. She was then referred for HROM, which demonstrated pan-oesophageal pressurisation with non-relaxing lower oesophageal sphincter: Type II Achalasia.

Achalasia in patients with OA/TOF is rare. We recommend inclusion of HROM in addition to TBE in children with OA/TOF having refractory reflux symptoms. Further research is required to evaluate the aetiology of oesophageal dysmotility in patients with OA/TOF.

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OC70

Unsafe swallow or eosinophilic oesophagitis (EoE), the importance of the multidisciplinary team for progressing nutrition.

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A 2-year-old female was referred to a paediatric gastroenterology clinic for the consideration of gastrostomy due to an unsafe swallow and recurrent chest infections. She had no other atopy. Other past medical history includes global developmental delay, previous subglottic stenosis and previous obstructive sleep apnoea.

Video fluoroscopic Swallow Study (VFSS) confirmed oropharyngeal dysphagia, characterised by delayed swallow initiation, and reduced airway closure, suggesting a significant aspiration risk. At the time of referral, the patient was receiving 75% of her estimated average requirements (EAR) via NGT using a whole protein feed. Small amounts of pureed foods were tolerated orally, and all liquids were administered via the NGT.

Upper gastrointestinal (GI) contrast study revealed a long delay in the passage, suggesting suboptimal oesophageal motility. pH impedance testing was within normal limits. Endoscopy performed in June 2022, concurrent with gastrostomy placement, identified active eosinophilic esophagitis (EoE), with 46, 18 and 1 eosinophils per high power field (eos/hpf) in the proximal, mid and distal oesophagus respectively. The patient was started on proton pump inhibitor (PPI) therapy.

A repeat endoscopy in November 2022 demonstrated persistent active EoE (36, 22 and 14 eos/hpf in the proximal, mid and distal oesophagus respectively). The patient's oral intake had improved, with greater consumption of soft-textured foods, but continued unsafe swallow for liquids. Gastrostomy feeds were reduced to 50% of EAR. A milk- and wheat-free diet (2 food exclusion diet [2FED]) was initiated to manage EoE as per BSG/BSPGHAN guidelines (2022), and the gastrostomy feed was changed to an amino acid formula (AAF).

By May 2023, repeat endoscopy showed EoE in histological remission on the 2FED. EAR was met via oral intake of family meals of varying textures, and gastrostomy feeds were discontinued. Liquids continued to be administered via the gastrostomy; a repeat VFSS was planned. Wheat was reintroduced into the diet, while a dairy-free diet continued.

In October 2023, endoscopy confirmed continued histological remission of EoE. The patient was advised to transition to a fully unrestricted diet, no feeds were administered via the gastrostomy, although this was still required for liquids. By April 2024, no active EoE was detected, although a slight increase in eosinophil count (4 eos/hpf in the proximal oesophagus) was noted. The patient achieved full oral nutrition and hydration, with the gastrostomy used exclusively for medication. Repeat VFSS indicated significant improvement in swallow physiology, with only mild residual difficulties in timing and coordination.

This case highlights the complex interplay between oropharyngeal dysphagia and EoE in paediatric patients. With a MDT approach, the patient achieved histological remission of EoE and significant improvement in swallowing function, leading to the eventual cessation of gastrostomy feeding and return to an unrestricted diet. This case underscores the importance of tailored, long-term management strategies in paediatric patients with coexisting gastrointestinal and swallowing disorders.

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OC71

Clabsi in Paediatric Home PN Patients

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Central Line-Associated Bloodstream Infections (CLABSI) represent a critical challenge in paediatric care, particularly for patients requiring home parenteral nutrition (PN). CLABSI refers to bloodstream infections seen in patients with central venous catheters (CVC), which are commonly used to administer intravenous fluids, medications, and nutrition when oral or enteral feeding is not feasible. In paediatric populations, especially those with chronic conditions or complex nutritional needs, central venous access is indispensable for the provision of home PN. However, the presence of these catheters significantly increases the risk of infection, particularly CLABSI, making it an ongoing concern for clinicians managing this vulnerable group.

We aimed to analyse the distribution of CLABSI episodes, the time to antibiotic administration, fever episodes, non CLABSI diagnosis, and the microbiological organisms associated with the infections. Additionally, the impact of different screening protocols on CLABSI management and outcomes is examined.

This study investigates the epidemiology and management of CLABSI in a cohort of 37 patients on home parenteral nutrition over the past 2 years. Data was collected on age, diagnosis, how many times a week their lines are accessed, episode of fever, causative microorganisms, screening and time to antibiotics for CLABSI episodes.

26/37 patients (70%) presented with fever at least once during the study period. Age at presentation ranged from 1 – 16. 31/37 (84%) used Taurolock as line lock, 3/31 (8%) used Tarolidine and 3/37 (8%) used heparin. Diagnoses included surgical short bowel syndrome, enteropathy, chronic intestinal pseudo-obstruction, trichohepatoenteric syndrome, cystic fibrosis, DGAT Mutation, MYO5B gene mutation and small bowel sclerosis.

There were 102 episodes of fever, median of n=2 per patient. 22/102 (21.5%) being due to CLABSI. 15.7% had another diagnosis like viral URTI (Rhinovirus (n=7) and Influenza (n=5) were most common) UTI, viral enteritis, and 62.75% with no causative organism isolated.

A small proportion (n=4) of patients experienced multiple episodes of CLABSI, with the highest number of episodes being four in a single case.

Enterococcus faecium (n=4 was the most frequently isolated pathogen from central line culture, followed by Staphylococcus epidermidis (n=3), Staphylococcus aureus (n=3), and Candida (n=3). Time from decision to give to administration of antibiotics was collected for the 22 episodes of CLABSI. 11/22 (50%) received antibiotics within 0-1 hour of decision, while 8/22 (36.36%) received antibiotics within 1-2 hours, and 3(13.64%) received antibiotics >3 hours after the decision to start antibiotics was made.

The majority of fevers in our CVC patient population were not associated with CLABSI. The majority of CLABSI cases were single episodes, but a small proportion of patients experienced recurrent CLABSI. All cases of suspected CLABSI should receive IV antibiotics within 1 hour of decision to treat; barriers to this are under review. Candida species has been implicated (n=4, 18% of CLABSI), a high index of suspicion should be maintained for this.

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OC72

Five-year review of patients with central venous access receiving home parenteral nutrition , who underwent endoscopies with antibiotic prophylaxis and analysis of post endoscopy infection rates at our tertiary centre.

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There are no established guidelines for antibiotic prophylaxis in children with central venous catheters (CVCs) on home parenteral nutrition (HPN), leading to varying practices across UK Centres. We hypothesize that children with intestinal failure are at increased risk for bacteraemia due to altered anatomy, dysmotility, inflammation, biofilm formation in long-term CVCs, and the use of central lines during procedures. Given the bacteraemia rates of up to 8% in upper and 25% in lower endoscopy for adults without central lines, we argue that prophylactic antibiotics are reasonable, given the increased risks faced by this high-risk group of children. We conducted a five-year review of patients with central venous access receiving home parenteral nutrition (HPN) who underwent endoscopies with antibiotic prophylaxis at our center (tertiary). We documented and analyzed post-procedure infections and their associated risk factors.

A total 48 patients receive home parenteral nutrition at our centre , 41 were analysed, 29 had antibiotics, 15 did not,receive antibiotics.

These 15 patients underwent 29 endoscopic procedures, including 4 upper, 9 combined upper and lower, and 16 combined upper, lower, and ileoscopy. Confirmed infection rates remained at 0% up to 28 days post-procedure. The agreed-upon prophylaxis regimen was implemented, with ciprofloxacin and metronidazole administered as the primary antibiotics. Notably, only 51.7% of patients received a peripheral cannula despite recommendations to avoid central line use during anesthesia, and 20.6% had small intestinal bacterial overgrowth.This study is the first to investigate post-endoscopy infection rates in pediatric patients on HPN. Despite a small sample size, we observed a 0% infection rate, significantly lower than reported rates in adults. These findings suggest that further research is warranted to explore the implications of antibiotic prophylaxis in this unique patient cohort and to establish guidelines that may enhance patient safety during endoscopic procedures

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OC73

Pneumatosis Intestinalis in Paediatric Patients with Neurodisability: A Case Series

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Patients with neurodisability can lack the capacity to communicate their symptoms. Understanding the natural history and implications of Pneumatosis Intestinalis (PI) in this group of patients could help refine diagnostic and therapeutic strategies, potentially sparing unnecessary surgical interventions. A retrospective keyword search of a radiology database identified patients diagnosed with PI via imaging over a 10 year period (2014-2024). Exclusion criteria were applied, selecting paediatric patients 6 months to 16 years of age with PI and neurodisability. Patient notes were analysed to identify: 1) subtle or atypical signs that may be present despite lack of clear symptoms 2) patients management 3) patient outcomes in form of clinical resolution and recurrence of PI. Five patients were identified (Table 1).

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Demographics: - Age at finding -Sex (M – male, F – female)	6 years M	7 months F	1 year 5 months F	3 years M	11 years F
Feeding Method	Gastrostomy	Nasojejunal	Gastrostomy	Gastrostomy Jejunostomy	Jejunostomy
Neurological diagnosis	Pompe disease	Undiagnosed neurological disorder with dystonia Severe development delay	Microcephaly Severe development delay	Epileptic encephalopathy Central hypotonia, muscle weakness, severe development delay	Mitochondrial depletion (SERAC1 mutation) syndrome Profound Learning Disability Epilepsy
Clinical presentation	Respiratory failure	Drowsy, less responsive hypothermic, hypotonic, Abdominal distention	Bronchiolitis Abdomen distended, Per rectum bleeding	Increased loose stool, bloating, evidence of discomfort	Asymptomatic Blood in stool 2 weeks prior
Imaging	Bubbly lucencies below diaphragm	Distended abdomen with several loops of dilated gas-filled bowel. Bubbly lucencies within left flank	Intramural gas within the bowel in the upper left quadrant	Extensive bowel wall thickening with intramural gas	Extremely severe and widespread pneumatosis
Pneumoperitoneum Present?	No	No	No	No	Yes (on 2nd admission)
Management (NBM - nil by mouth, Abx - antibiotics, PN - parenteral nutrition)	Treated for respiratory failure with 1 week Tazocin Conservative management for Pneumatosis	NBM + Metronidazole	IV abx + NBM + PN	Abx + NBM	Abx + NBM + PN
PI duration	32 days	17 days	50 days	5 days	487 days (ongoing)
Recurrent Episodes (PI on separate admission)	0	0	0	0	2

All 5 patients were managed conservatively, with all but one having resolution of PI noted on imaging. In the absence of any specific surgical intervention, only one patient experienced recurrent PI. Despite this, the patient remained asymptomatic from a gastrointestinal perspective on each subsequent episode. These cases demonstrate that a conservative approach to management can be effective and should not be ruled out in favour of surgical management.

OC74

Characteristics and management of upper gastrointestinal bleeding (UGIB) in a tertiary paediatric gastroenterology centre: a retrospective review

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UGIB is a potentially life-threatening event, defined as bleeding from above the fourth part of the duodenum manifesting as haematemesis, coffee-ground vomiting or melaena (1). It is rare in the paediatric population with causes varying by age. Many guidelines are extrapolated from adult guidelines (2,3). BSPGHAN produced a regional pathway for UGIB in 2018 (4). This review describes aetiology and management of UGIB in children in a single tertiary centre across an eighteen-month period.

Data was gathered by searching electronic patient discharges for coded diagnoses of 'upper gastrointestinal bleed', 'upper gastrointestinal haemorrhage', 'haematemesis', 'coffee ground vomit' or 'melaena'. Handover lists were also analysed. Patients aged sixteen or under with signs of UGIB were included; those with only LGIB (fresh rectal bleeding) were excluded.

35 children were identified. 15/35 presented to the tertiary centre emergency department, 15/35 were transferred from district general hospitals (DGH), 4/35 were current inpatients or day-case patients and 1/35 was undocumented. 21/35 had significant co-morbidities; 9/21 had known GI conditions. 17/35 presented with minor bleeding without haemoglobin drop; 18/35 had significant bleeding with haemoglobin drop or low haemoglobin on admission requiring transfusion.

The majority had documented A to E assessments and were kept nil by mouth but investigations performed differed. Most children (33/35) did not have a chest x-ray or documented consideration of button battery ingestion. 12 did not have a full blood work up, with clotting studies most commonly missing (7/12). Most received intravenous (IV) omeprazole and 10/35 received IV tranexamic acid. 11/35 were given IV antibiotics without documentation of indication.

19/35 underwent endoscopy, 7/35 are awaiting endoscopy, 4/35 had other causes identified and 5/35 had minor bleeding and were discharged without follow up. 3/35 patients required endoscopic intervention including adrenaline, polypectomy and flouracil with balloon tamponade.
Causes identified for UGIB are summarised in table one.

Significant GI pathology	8	
Liver disease with portal hypertension	1	
In context of other acute illness	2	
Post endoscopy	2	
Irritation from gastro-jejunal tube	2	
Gastritis	H pylori positive	H pylori negative
	2	8
Mallory Weiss tear	4	
Other	6	

Table one – causes for UGIB

Significant GI pathology included a gastrointestinal stromal tumour (1), Meckel's diverticulum (2), Peutz-Jegher polyposis (1), gastric ulcer (1), duodenal ulcer (2), severe oesophagitis (1). 'Other' included swallowed maternal blood, self-induced, and minor bleeds.

Limitations of this study were the retrospective nature and small numbers. The presentation and causes for UGIB in paediatric patients remains broad. Although rare, significant GI pathology should remain in the differential. Around half presented with significant bleeding requiring transfusion. However, since the need for endoscopic intervention is uncommon, adult Gastroenterology and surgeons may be required to support endoscopic therapy.

Numerous presentations at DGHs show education and network management is vital to support paediatricians in the stabilisation and management of UGIB. Although no patients had ingested a button battery, documentation of this consideration was inadequate. This is vital due to the associated morbidity and mortality. Documentation of clinical reasoning for utilising different medications for UGIB remains an issue, within and without gastroenterology.

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OC75

Interferential therapy for treatment-resistant symptoms of GI dystonias: A case series

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Gastrointestinal dystonia (GID) in children with neurological impairment present with symptoms such as pain, retching, and vomiting linked to feeding and bowel habits. Interferential therapy (IFT), a low-frequency electrical treatment traditionally used for muscle pain, was applied in three resistant cases of GID, showing potential in alleviating gastrointestinal pain and distress. A 4-year-old with Phelan McDermid syndrome, developmental delay, and hydrocephalus presented with vomiting, crying, and constipation which was unresponsive to pharmacological treatments. After initiating IFT alongside existing therapies, the child experienced reduced pain, vomiting, and constipation, allowing discontinuation of some medications, with sustained improvements in bowel regularity and nutritional status. Another 13-year-old with brainstem ganglioglioma suffered from progressive neurological and autonomic dysfunction, including vomiting, unsafe swallowing, and bowel irregularities, despite prior surgical and medical interventions. IFT improved bowel function, PEG-J feed tolerance, and enabled discontinuation of parenteral nutrition, although recent abdominal pain required further medical attention. A 12-year-old with Trisomy 12 mosaic, GERD, and chronic gastrointestinal issues, including abdominal distention and constipation, showed initial improvements in bowel function and enteral feed tolerance after starting IFT. However, symptoms returned after three months, leading to the cessation of IFT and resumption of parenteral nutrition, with motility studies revealing small bowel neuropathy and colonic inertia. This case series underscores the potential of IFT in managing GID in pediatric patients with complex neurological conditions, demonstrating symptom improvement without adverse effects. While responses varied, further research into IFT's efficacy and limitations is recommended.

OC76

Oesophageal Strictures in a Paediatric Tertiary Care Centre: A 14-year Experience.

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Oesophageal strictures in the paediatric population pose a significant clinical challenge, with affected children often requiring recurrent endoscopic dilations to manage their condition [1]. This retrospective study aimed to analyse the demographics, aetiologies, management, and outcomes of children treated for oesophageal strictures at a tertiary care centre in London over a 14-year period. We reviewed medical records of patients aged 18 years or younger who underwent oesophageal dilatation for benign oesophageal strictures between January 2010 and September 2024. Data collected included patient demographics, stricture aetiology, number of dilatations, need for adjunctive interventions (e.g., stent placement, intralesional steroid injections or topical mitomycin application), and treatment outcomes.

Our findings revealed that anastomotic strictures were the most prevalent (39/55, 71%), followed by corrosive (4/55, 7.3%), and peptic strictures (4/55, 7.3%).

Endoscopic balloon dilatation was demonstrated to be a highly effective primary treatment approach, achieving successful outcomes in the majority of cases (36/55, 65.45%).

Additionally, three patients with successful treatment outcomes necessitated a combination of dilatation and stent placement, while two others required dilatation in conjunction with medication, and one patient necessitated dilatation, medication, and stent insertion to attain a satisfactory result. Notably, patients with caustic strictures presented significant management challenges, often necessitating multiple dilatations (ranging from 15 to 56) and adjunctive interventions such as stent placement, steroid injections and mitomycin application.

Of the 55 patients included in the study, 15 (27%) had evidence of dysmotility on barium swallow, and 13 (23.6%) had gastroesophageal reflux disease confirmed by pH/impedance testing.

Furthermore, our study found that eosinophilic oesophagitis was a contributing factor to oesophageal dysmotility in 3 patients (5.45%), all of whom had an underlying aetiology of anastomotic strictures following repair of oesophageal atresia with tracheoesophageal fistula. This observation holds clinical significance, as eosinophilic esophagitis can induce fibrosis and structural changes within the oesophagus, potentially exacerbating the severity and recurrence of oesophageal strictures.

Successful long-term management of oesophageal strictures in children relies on the careful assessment and management of co-existing conditions, such as gastroesophageal reflux disease and oesophageal dysmotility, which are significant co-morbidities in this patient population [2] .This study underscores the complexity and multifaceted nature of oesophageal strictures in the paediatric population, emphasizing the need for a multidisciplinary approach to optimize management strategies and long-term outcomes.

OC77

Exclusive enteral nutrition for induction of remission in paediatric Crohn's disease: A single centre experience

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Exclusive enteral nutrition (EEN) is recommended by the European and North American Societies for Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN, NASPGHAN) as first line remission induction in paediatric Crohn's disease. Remission is induced in up to 80%[1]. Advantages include provision of a nutritionally complete feed and minimal side effects compared to corticosteroids[2]. Proposed mechanisms include effect on intestinal immune function, direct anti-inflammatory effects and induction of microbiome changes[1,2,3]. Whole protein feed is recommended, literature is limited on individual products.
We aimed to evaluate remission rates and outcomes of Ensure Plus/Paediasure Plus as EEN in our tertiary centre.

Patients receiving EEN for induction of remission (first occurrence) were identified from the departmental database. Data collected included demographics, disease characteristics, admission route, refeeding risk, days to target volume, prolonged stay, completion and remission rates, further induction and maintenance treatment, ongoing supplementation.

50 patients were identified over a 12-month period. Mean age at diagnosis was 11.58 years. Disease distribution: 2/50 ileal, 15/50 ileocolonic, 17/50 upper gastrointestinal (UGI) + ileocolonic, 4/50 colonic + UGI, 12/50 colonic only. Further subtyping included 35/50 luminal, 12/50 fistulating, 1/50 fistulating + stricturing, 2/50 stricturing, 13/50 perianal.

Percentage median BMI (%mBMI) was >80 in 37/50, 70-80 in 7/50, <70 in 2/50. All with %mBMI <80 had ileal or UGI disease. 19/50 were high refeeding risk, based on %mBMI, oral intake, weight loss.

41/50 used Ensure Plus, 8/50 Paediasure Plus, 1/50 EO28 (milk allergy).

37/50 completed orally, 11/50 via nasogastric tube (NGT), 2/50 combination. 49/50 were admitted to commence EEN, 1/50 did home EEN. 23/49 were admitted from theatre, 15/49 electively, 3/49 via Emergency Department (ED), 6/49 transferred from a district general hospital (DGH).

Modal days to target volume was 3 (range 2 – 9). Modal length of stay was 4 days (range 2 – 42). 19/49 had a prolonged stay: 11/49 nutritional rehabilitation prior to endoscopy, 4/49 NGT training, 1/49 to meet target, 3/49 for other management.

9/49 required oral phosphate replacement during EEN, 1/49 required oral magnesium. No patients required IV replacement. 9/10 (90%) requiring electrolyte replacement were identified as high refeeding risk.

38/50 (76%) completed EEN. Reasons for discontinuation included: no improvement at 4 weeks (7/50), vomiting (2/50), erythema nodosum (2/50), patient choice (1/50). 12/50 (24%) continue on nutritional supplements.

22/50 were excluded from further analysis (top-down treatment 9/22; early cessation 12/22, alternative feed 1/22).

28/50 were analysed further. 26/28 demonstrated weight gain. 26/28 were in clinical remission at 8 weeks, 24/28 at 3 months, 19/28 at 6 months, 15/20 at 12 months. 3/28 required steroids, 2/28 second EEN course, 2/28 steroids and EEN. 3/28 later commenced biologics.

Ensure/Paediasure Plus is well tolerated and effective. Most complete EEN orally with average 3 days to target volume. Remission rates are high at EEN completion and 3 months (93%, 86%). 68% and 75% respectively sustained remission at 6 and 12 months. A minority required further EEN or steroid courses or later biologic therapy. None required IV electrolyte replacement regardless of initial refeeding risk category. Home EEN may be safe and effective.

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OC78

Cutaneous Skin Manifestations Associated with TNF Alpha Inhibitor in Paediatric Patients with Inflammatory Bowel Disease: A Case Series from a District General Hospital in the United Kingdom

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Anti-TNF-alpha therapies are effective for treating inflammatory bowel disease (IBD) but can be associated with dermatologic complications, including infections, psoriasis, and eczema. While there are reports of anti-TNF-related skin reactions in IBD, data specific to paediatric populations are limited. Recognizing and managing these skin manifestations early is essential to optimize care for this population.
We conducted a retrospective review of six paediatric patients with IBD who developed anti-TNF-induced skin manifestations. Collected data included patient demographics, IBD phenotype, type of dermatologic reaction, anti-TNF dosing, clinical interventions, and outcomes.

The patients, aged 8-14 years, included an equal number of females and males. Five (83%) had Crohn's disease, and one (17%) had ulcerative colitis; all were on infliximab at the time their skin reactions developed. Cutaneous reactions included psoriatic lesions on the knees, elbows, legs, and scalp (1 patient); lesions on the ears (3 patients); nostril involvement (2 patients); scalp lesions (2 patients); and inner breast area lesions (1 patient). These findings illustrate the broad range of possible skin manifestations, which can vary significantly in distribution and severity.

All patients were receiving infliximab with dosing ranging from 5 mg/kg to 10 mg/kg and infusion intervals between 4 to 8 weeks. Infliximab trough levels were typically below 10 mg/mL though one patient had levels exceeded 15 mg/mL. Most lesions were identified as inflammatory or infective, with one case diagnosed as psoriasis. Treatment included topical steroids (mometasone, clobetasone) in 90% of cases, and all patients received antibiotics at the initial stage of skin involvement. Only one patient required discontinuation of infliximab.

Our findings suggest that paediatric IBD patients can develop anti-TNF-induced skin reactions without a personal or family history of dermatologic conditions, posing a diagnostic and management challenge. Importantly, these reactions may appear even when IBD is well-controlled, making decisions regarding continuation or cessation of therapy more complex.

Physicians should maintain a high index of suspicion for anti-TNF-induced dermatologic manifestations in paediatric IBD patients who present with new or unusual skin lesions. Assessing infliximab trough levels may help in evaluating whether higher concentrations contribute to skin reactions, though further research is needed to establish a definitive link. Clinicians should consider early dermatologic consultation and weigh the benefits and risks of continuing anti-TNF therapy considering these potential adverse effects, particularly when IBD control is stable but dermatologic complications arise.

By identifying and managing these skin reactions proactively, healthcare providers can better support the overall therapeutic goals for paediatric IBD patients, optimizing treatment outcomes and patient quality of life.

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OC79

SAPHO Syndrome in a Child with Crohn's Disease: A Case Report

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SAPHO (Synovitis Acne Pustulosis Hyperostosis and Osteitis) syndrome is a rare autoinflammatory condition characterized by bone inflammation (aseptic osteitis) and skin manifestations, such as palmoplantar pustulosis, acne, and pustular psoriasis. Due to its often subtle and variable presentation, SAPHO syndrome is frequently misdiagnosed, especially in patients with comorbid inflammatory bowel disease (IBD). Since 1992, cases of SAPHO syndrome associated with IBD have been reported, though only a few paediatric cases have been documented. Here, we present a unique case of SAPHO syndrome developing in an adolescent with Crohn's disease, highlighting the diagnostic and therapeutic challenges associated with overlapping autoimmune diseases.

An 8-year-old girl was seen in the paediatric Gastroenterology clinic with symptoms of chronic diarrhoea, angular stomatitis. Her Faecal calprotectin was >1800. Endoscopy and histology revealed multifocal Crohn's disease-Stomach, Duodenum, ileum and Colon (Paris Classification A1b, L3 + L4a, B1, G0)

After diagnosis, the patient achieved remission with Modulen and corticosteroids. However, maintenance therapy with azathioprine proved ineffective in maintaining clinical remission and Infliximab was initiated. Due to ongoing symptoms her IFX dose was increased to 10 mg/kg every 8 weeks, and azathioprine was discontinued. She maintained sustained clinical and biochemical remission after optimising the IFX therapy.

At age 11, three years after her Crohn's diagnosis, she developed psoriatic skin lesions, including palmar psoriasis and plaques on her knees, elbows, scalp, and legs. Dermatology initially suspected an anti-TNF-induced skin reaction; however, the diagnosis was revised to psoriasis, and topical therapy with steroid and emollients was started. Around the same period, she developed painful swelling at the medial end of the right clavicle, presenting with warmth, mild tenderness, and persistent erythema. Radiological studies revealed chronic recurrent multifocal osteomyelitis (CRMO) of the right sternoclavicular joint, fulfilling the Kahn criteria for a diagnosis of SAPHO syndrome.

Due to her stable Crohn's disease and the onset of SAPHO-related symptoms, infliximab was discontinued at age 12. However, as pain in her clavicles, knee, and elbow intensified, she received a Zolendronic acid infusion for symptom relief. With her Crohn's disease in remission, she was switched to weekly subcutaneous methotrexate (15 mg/m^2).

Over the past two years, the patient has maintained a stable clinical course, with sustained clinical and biochemical remission of her Crohn's disease and complete resolution of osteoarticular and dermatological manifestations.

This case underscores the complexity of diagnosing and managing SAPHO syndrome in paediatric patients with Crohn's disease. SAPHO syndrome can mimic anti-TNF skin reactions, adding diagnostic challenges in IBD patients. Methotrexate proved effective in managing SAPHO in this patient and may offer an effective alternative therapy allowing for sustained remission of both Crohn's disease and SAPHO symptoms. Biologic swap could be considered to Adalimumab if Methotrexate alone does not provide adequate maintenance therapy for Crohn's. Multidisciplinary collaboration and individualized therapy are essential in managing the intersecting pathways of autoinflammatory and autoimmune conditions in paediatric patients.

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OC80

Clinical Audit and Quality Improvement: Faecal Calprotectin Measurement in Paediatric IBD Outpatient Clinics

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Faecal calprotectin (FC) is an established biomarker used for monitoring bowel inflammation in paediatric inflammatory bowel diseases (PIBD). ESPGHAN IBD guidelines recommend measurement of FC 6-monthly(1–3). Additionally, normalisation of FC is a treatment target per STRIDE II.(4)

Aim: report frequency of FC sample measurement in PIBD outpatient setting and whether frequency of FC processing in patients diagnosed with IBD ≥1 year meets the 6-monthly target.

Method: Retrospective chart review of PIBD patients attending our tertiary centre over six months between 1/1/23-30/6/23. Demographics, diagnosis, duration of diagnosis, FC measurement at clinic visit, interval between FC measurements were recorded. We reported on individual clinic visits and patients diagnosed ≥1year.

Results: Total 385 clinic contacts. 218 included for analysis, representing 176 unique patients. 54.5%(n=96) male. 48.8%(n=86) CD. 45.5%(n=80) UC. 5.7%(n=10) IBDU. Median [IQR] age 14 years [11 to 15], duration of diagnosis: 2 years [0.9 to 4.2].

Results in Table 1:

Table 1: Results

Outcome	**Result %(n)**
All patients (n=176)	
FC processed for clinic contact	39.7(87)
Patients with diagnosis ≥1year (n=134)	
≥2 FC processed ≥2 FC processed, interval ≤6 months Median interval between samples: 8 months [4.7 to12.3]	 64.9(87) 26.1(35)
1 FC processed	28.4(38)
0 FC processed	6.7(9)

	Patients with ≥ 2 FC	**Patients with 1 FC**	**Patients with no FC**
n(male)	114(62)	50(27)	13(8)
Age Years, Median [IQR]	13[10 to 15]	14[13 to 15]	13[12 to 16]
Diagnosis	CD: 50.8 UC: 41.2% IBDU: 7.9%	CD: 44% UC: 54% IBDU: 2%	CD: 53.9% UC: 46.1%
Duration of diagnosis Years, Median [IQR]	1.9[0.8 to 4]	2.1 [1 to 4.1]	1.7[0.9 to 5.7]
Inter-sample interval Months, Median [IQR]	7.1[4 to 11.1]	N/A	N/A

Only 39.7% of PIBD clinic visits have an associated FC result and we only meet the standard of measuring FC 6-monthly in 26.1% of patients with established diagnoses of IBD ≥1 year. Our processing rate is lower than the currently published data for FC compliance in both children and adults ranging from 66.7-78%.(5,6) While our study utilises a large number of patients and a representative spread of diagnoses, a limitation is that we did not investigate reasons for low rates of FC measurement. We suggest the following likely reasons: patients unable to open bowels in clinic on-demand; difficulty obtaining samples from patients living geographically distant to our site; difficulty obtaining results from samples processed in primary care.

We introduced the following interventions:

1. Modification of clinic invitation to include reminders to bring stool samples to clinic.
2. SMS reminders the day before clinic to encourage patients to bring stool samples.
3. Patient-lead postal stool samples. This does have financial implications for the patient and trust, in the form of postage and packaging costs to ensure biological substance transport compliance.

We suggest these simple technological solutions such as direct messaging patients, and offer postal stool sampling as a safe, effective failsafe. Re-audit post-implementation of interventions is due in 2025.

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OC81

Frequency of biologic switching and outcomes of multiple biologics switching in children and young people with Inflammatory bowel disease: Major centre experience

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Information is limited about biologics switching pattern in children and young people (age <= 18 years) with inflammatory bowel disease (IBD) in an era of many biologics therapies. The best choice of biologic to use if the first biological infliximab is not beneficial is also unclear. This retrospective observational cohort study aimed to quantify and describe biologics switching patterns in children and young people with IBD, and to compare the effectiveness of using a second tumor necrosis factor inhibitor (TNFi) versus non-TNF failure of a first biologic TNFi in routine clinical practice.

Patients with two or more successive biologic switches were identified. Biologic treatment sequence was analyzed descriptively. Baseline demographic and clinical characteristics were collected retrospectively from the electronic medical records. Treatment failure was defined as the composite of IBD related hospitalization, new prescriptions of oral/ IV corticosteroids or any IBD related surgery or the need to switch to a third biologic agent. We compare characteristics of early switchers (<=6 months) vs late switchers (> 6 months).

Total 229 patients were identified whom received tumor necrosis factor a antagonist (anti TNF a) treatment followed by another anti-TNFa treatment or Vedolizumab then Ustekinumab. Median age at diagnosis was 8 years. We found that (40 %) switched to a different biologic after a median of 2 years after diagnosis. In total thirty six of total 229(15%)percent of patients received 3 or more biologics. The most common reason for switching from antiTNF was poor clinical response in (50%) followed by antibodies and one patient was switched due to difficult access. Seventy five percent were late switchers, and switched primarily due to loss of response or antibody formation.

Most patients who failed vedolizumab had either ileocolonic disease, perianal disease or a combination. Median duration of vedolizumab was 1 year.

3 of total 229 patients with ileocolonic fistulating phenotype showed poor response to Ustekinumab and required IBD related surgery. One patient was subsequently switched to Risankizumab (awaiting evaluation) with 1 further patient for consideration of risankizumab.

Vedolizumab was the most common second line biologic regardless of phenotype; patients with ileocolonic/perianal disease showed poor response. Ustekinumab may be more suitable as second line in ileocolonic or perianal disease. Large multicentre studies will be beneficial as newer agents become available in paediatric IBD.

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OC82

The diagnostic pathway of paediatric inflammatory bowel disease in a Scottish regional centre before and after the Covid pandemic: Good access to endoscopy but MRI lagging behind

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Health Services nationwide have faced challenges following the COVID-19 pandemic. The aim of this review was to review our current practices against the IBD UK Standards and how this has changed following the COVID-19 pandemic.

Patients diagnosed with Paediatric Inflammatory Bowel Disease (PIBD) within a tertiary gastroenterology centre in Scotland from 1/1/18 to 31/12/23 were identified using an existing prospective IBD patient register. Patients were divided into Pre-Covid (diagnosis before 23/3/20), and Post-Covid (diagnosis from 19/7/21) epochs, with the interval Covid epoch (24/3/20-18/7/21 inclusive) also presented for interest. Electronic patient records were reviewed to identify time from referral to gastroenterology review, and time from request to endoscopic assessment and small bowel MRI. These outcomes were compared to IBD standards statements 2.2 (that patients referred with suspected IBD should be reviewed within 4 weeks) and 1.10 (that endoscopy and MRI should be accessible within 4 weeks).

123 patients were identified as having been diagnosed with IBD. 35 Pre, 66 Post and 22 Covid, and are summarised in Table 1.

	Total	Pre-Covid	Covid	Post-Covid
Patients	123	35	22	66
Median age at diagnosis (years)	13.11 (3.54-17.26)	12.90 (3.54-16.68)	13.54 (6.44-17.26)	13.45 (3.85-16.98)
Diagnosis:				
Crohn's Disease	68 (55.3%)	18 (51.4%)	13 (59.1%)	37 (56.1%)
Ulcerative colitis	44 (35.8%)	12 (34.3%)	7 (31.8%)	25 (37.9%)
IBDU	11 (8.9%)	5 (14.3%)	2 (9.1%)	4 (6.0%)
Source of referral:				
Inpatient	27 (22.0%)	2 (5.7%)	5 (22.7%)	20 (30.3%)
Outpatient	88 (71.5%)	32 (91.4%)	14 (63.6%)	42 (63.6%)
Not documented	8 (6.5%)	1 (2.9%)	3 (13.6%)	4 (6.1%)

Table 1 – Demographics of PIBD patients diagnosed 01/01/18 – 31/12/23, and split per diagnostic epoch

In the pre cohort, 70.4% (19/27) patients with documented referral and tertiary outpatient review were seen within 28 days. This has fallen following the pandemic with 65.8% (25/38) patients meeting standards. There was a non-significant drop in median wait time for tertiary outpatient review from pre- (16 days (range 1-82 days)) compared to post-Covid cohorts (9 days (range 0 –274 days, p=0.06).

71.4% (25/35) patients in the pre-covid cohort underwent endoscopic evaluation within 28 days. Median wait time was 15 days (Range 1-63 days). The proportion of patients within target for undergoing endoscopic assessment rose to 91.9% (57/62) patients in the post-COVID cohort. The median wait reduced to 6 days (range 1-112 days, p=0.008).

Small bowel MRI within 28 days of request was achieved in a similar proportion of both the Pre (18.2%; 6/33) and Post cohorts (18.3%; 11/60). However, median time to MRI was significantly higher in the Post cohort (111.5 days; range 4-303 days) compared to the Pre cohort (56 days; range 4-227 days, p=0.03).

Despite the NHS-wide impact of the COVID-19 pandemic, this service audit has shown stability or improvement in waiting times for both tertiary review and subsequent endoscopic assessment in those with suspected IBD. Access to MRI remains problematic and has considerably worsened after COVID-19, with median wait more than doubling compared to Pre-Covid. More data regarding this nationwide would be welcome to understand and modify the delays seen here.

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OC83

A 5 year single centre clinical practice review of the safety and efficacy of Ustekinumab treatment 2019-2024 for children with refractory IBD.

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Ustekinumab is a monoclonal antibody targeting interleukin 12 and interleukin 23 pathways in the treatment of IBD, widely used in both adult and paediatric populations with reported safety and efficacy in achieving clinical remission[1,2]. This review was prompted by 3 significant adverse reactions requiring resuscitation with 2/3 children receiving IM Adrenaline for suspected anaphylaxis.

The prospective local data base of all children diagnosed with IBD 2019-24, total 450, was reviewed and those treated with Ustekinumab were included in the study. 32 patients were prescribed Ustekinumab. 20/32 were males. Age at start of treatment was 8.75-17.75 years and time from diagnosis to starting Ustekinumab was 5 months-9.25 years, mean of 3.6 years. 26/32 patients were diagnosed with Crohn's disease, 4 with ulcerative colitis, 1 with VEOIBD and 1 with IBDU. All patients had received prior, optimised, anti TNF treatment, 17 had received both Infliximab and Adalimumab. 9 patients had also received Vedolizumab (alpha 4 beta 7 integrin uptake blocker), and one Tofacitinib (JAK inhibitor).

At the time of data collection 14 children (43.75%) remained on Ustekinumab and of those 11 (34.37%) had been on it for 6 months or longer.

3 children experienced anaphylactoid reactions to the initial intravenous loading dose, 2 received IM adrenaline and oxygen. Serum tryptase was not raised in any of the cases. Although tryptase is commonly raised in anaphylaxis this is not always the case. These children switched therapies immediately.

Data for calprotectin levels was available for 27 children. 92% children had a calprotectin >200 at the beginning of treatment, with 63% >800. At three months 60% of children had a reduction in calprotectin levels but over a third, 36% remained >800. Data available for 11 children remaining on Ustekinumab > 6 months showed that half (53%) were in probable mucosal remission with faecal calprotectin <200, with a third (33%) showing clinical improvement but ongoing mucosal inflammation, calprotectin >800.

9/29 children were clinically well when commencing Ustekinumab treatment. Medication change was because of high calprotectin results, MRE results and adverse skin reactions to anti TNF treatment. At 6 months 8/9 children remained on Ustekinumab, 5 had no clinical symptoms, but 3 patients reported worsening symptoms. 5/6 children swapped due to adverse skin reactions to anti TNF reported improvement.

20/29 children had significant IBD clinical symptoms when starting Ustekinumab. 16/20 (80%) saw some improvements at 3 months. At 6 months 8/20 (40%) reported improved symptoms and were still on Ustekinumab. 12/40 (60%) reported no improvement or deterioration in their original symptoms, or had stopped the medication.

7/29 (24%) children, lost response to Ustekinumab after making initial improvements, when the intervals between injections was extended children (4-8 weekly) showed signs of relapse. This may suggest that more frequent dosing and / or reloading with IV doses may improve outcomes[3]. Drug levels for Ustekinumab were not available at the centre.

It is important to be aware of potential rare but significant adverse events when selecting medication and counselling families.

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OC84

Does early successful Kasai leads to better transplant free survival?

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Biliary Atresia (BA) is managed by Kasai Portoenterostomy (KPE) and or a Liver Transplantation (LT). There is evidence and acceptance of the fact that KPE performed at a younger age provides better jaundice clearance. It is also accepted that if KPE does not work LT will be required within 1yr, in most cases. However, it has not been shown if there is a difference in the need and timing of LT in children who clear jaundice after undergoing an early successful KPE compared to those who undergo successful KPE at a late stage. We would like to present this aspect of our findings.

We conducted a retrospective study on prospectively collected data between 2004 and 2023, with follow-up to 2024.

Total number of patients were 210.
Group 1 (KPE by day 30) n-51. Group 2 (KPE after 30 days of life) n-159.
Gp 1 n-35 (68.62%) cleared jaundice and n-16 did not, compared to Gp 2, n-97 (61%) cleared jaundice while n-62 did not.
Amongst the patients who cleared jaundice in Gp 1, n-5 (14.7%) required LT. Mean time to transplant was 69.6 months (range 15 to 185.3 months). There were no deaths.
In Gp 2, n-2 patients died and n-19 (19.58%) have required LT. Mean time to transplant was 47 months (range 6 months to 147 months).

Conclusion
Patients undergoing KPE in the first month of life have a better rate of Jaundice clearance.
Children undergoing early successful KPE are less likely to need a subsequent transplant than those who have a late successful KPE.
Children undergoing early successful KPE requiring a LT are likely to need LT later than children who have a late successful KPE.
Preliminary analysis on a data set including both children, who did and did not clear jaundice revealed that duration of survival to death or transplant in children with BA who had late KPE is reduced to about 38% of BA children undergoing early KPE. This result is statistically significant.

OC85

The biochemical and nutritional effects of odevixibat in patients with Progressive familial intrahepatic cholestasis and Alagille syndrome

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Progressive familial intrahepatic cholestasis (PFIC) and Alagille syndrome (ALGS) are inherited disorders leading to cholestasis and subsequent hepatocellular damage potentially requiring a liver transplant. Odevixibat, an oral inhibitor of the ileal bile acid transporter, is being studied for its effectiveness in treating PFIC and ALGS.

This retrospective case series evaluates patients with PFIC and ALGS treated with odevixibat at a tertiary centre between May 2022- October 2024. Electronic case notes were reviewed at 0,6,12-month time points for nutritional and biochemical parameters (mid-upper arm circumference (MUAC), weight, height, alanine aminotransferase (ALT), gamma-glutamyl transferase (GGT), and bilirubin levels).

Six patients were on odevixibat. Four patients had PFIC (2 BSEP mutations, 1 MDR3 deficiency, and 1 FIC1 deficiency) and two patients had ALGS.

In the PFIC group (all female), the mean age was 4.25 (1-8 years). Median duration of treatment with odevixibat, was 11.75 months (2-29). The notes were assessed at 0 months for all four patients and 6 months for three patients (1 patient had a liver transplant before 6 months). Two patients underwent liver transplant both in the PFIC group (1 BSEP, 1 MDR3 deficiency) at 2.5 months and 6 months respectively of treatment.

In ALGS group (all male) mean age was 2.5 (2-3 years). Median duration of treatment was 14.5 (14-15) months.

Table 1: Median values of biochemical and nutritional parameters.

	PFIC		**ALGS**		
Months	**0**	**6**	**0**	**6**	**12**
MUAC (cm)	13.9	15.9	13.05	14.6	15.6
Weight (kg)	11.22	16.5	7.01	8.49	9.77
Height (cm)	84.5	105	68.4	76.1	80.25
ALT (<60iu/L)	268.5	89	109.5	114.5	128
GGT (<40iu/L)	31	24	499.5	704.5	542.5
Bilirubin(2-21umol/L)	51	129	170.5	120	115.5
Albumin(30-40g/L)	36	28	31.5	29.5	33

In the PFIC group, three patients required NG/PEG supplementation. Two patients received a medium-chain triglyceride (MCT) diet, receiving Liquigen and or Heparon Junior. They maintained same height and weight centiles at 6 months, with one requiring nasogastric NG/PEG feeding. One patient reached the 12-month assessment and was on NG/PEG feeding for 4 months and the MCT diet. She demonstrated an improvement in height and weight centiles, rising from <0.4 to the 0.4th percentile, with stable ALT, GGT, and albumin concentrations. Two patients underwent liver transplant, pre liver transplant 1 patient showed an improving height and weight centile whilst the other remained the same.

In the ALGS cohort, both patients showed improved nutritional status at 6 and 12 months. Both received Liquigen via NG/PEG. Height centiles remained the same however weight centiles improved. ALT levels slowly increased over time, while GGT levels rose initially then improved by the 12-month mark. Bilirubin levels improved and albumin levels improved by 12 months.

In summary, this single centre experience of the use of odevixibat, demonstrated that the ALGS group showed improved biochemical parameters. In the PFIC group, this was not seen as two patients required liver transplantation. In both groups, odevixibat has demonstrated to be a useful adjunct to improve nutritional status.

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OC86

The clinical spectrum of porto-sinusoidal vascular disease in paediatrics: a single centre review

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Porto-sinusoidal vascular disease (PSVD) is a condition which affects the portal venules and sinusoids in the absence of cirrhosis[1]. Knowledge about PSVD natural history is still limited[2,3,4].

We aimed to describe the characteristics of children affected with PSVD, as well as their histological findings and clinical outcomes.

We performed a retrospective review of children with native liver biopsy samples suggestive of PSVD from January 2019 to October 2024.

Forty-eight patients fit the diagnostic criteria of PSVD as described by Gottardi et al.[1] with 64.6% (n=31) having associated portal hypertension (PHT) and 35.4% (n=17) without PHT. The most common reason for referral to hepatology was isolated elevated transaminases in the non-PHT group, hypersplenism and portal hypertensive bleed in the PHT group (table 1). There were no significant differences between the histological features found on both groups, apart from a higher prevalence of perisinusoidal fibrosis in those with PHT (p=0.018).

The non-PHT group had a higher median ALT of 61 (IQR 35, 213) at presentation when compared to the PHT group (p=0.021). During the follow up period no significant adverse events were observed in the non-PHT group, while 2 children developed hepatopulmonary syndrome and 3 underwent liver transplantation in the PHT group. Of those with PHT, 16 had evidence of portal vein thrombosis (PVT) with cavernous transformation at presentation and 2 developed PVT during the follow up period.

In summary, PSVD is a condition with a variable phenotypic presentation and disease course. Those without PHT, mostly presenting with isolated elevated transaminases, appear to have a milder disease course, while those with PHT can have poor outcomes. Prospective studies are required to improve our understanding of the disease course and identify those at risk of progressing to PHT and developing adverse outcomes.

Baseline characteristics and outcomes of children with PSVD

	Total	Non-PHT	PHT
Number of patients, n (%)	48 (100)	17 (35.4)	31 (64.6)
Gender: male, n (%)	30 (62.5)	9 (52.9)	21 (67.7)
Median age in years at presentation (IQR)	5.0 (2.1, 11.6)	4.3 (1.0, 11.7)	6.0 (2.6, 11.1)
Median follow up in years (IQR)	2.4 (1.6, 5.3)	2.1 (1.6, 2.4)	3.6 (1.9, 5.6)
Associated conditions, n (%)	35 (72.9)	13 (76.5)	22 (71.0)
Reason for referral to hepatology, n (%)			
Isolated elevation in transaminases	12 (25.0)	12 (70.6)	0 (0)
Splenomegaly	4 (8.3)	0 (0)	4 (12.9)
Hypersplenism	12 (25.5)	0 (0)	12 (38.7)
Portal Hypertensive bleed	12 (25.5)	0 (0)	12 (38.7)
Other	8 (16.7)	5 (29.4)	3 (9.7)
Liver elastography, median in Kpa (IQR)	7.7 (6.0, 10.1)	6.9 (5.1, 7.7)	8.6 (6.1, 10.9)
Management, n (%)			
Conservative	17 (35.4)	17 (100)	0 (0)
Endoscopic surveillance	22 (45.8)	0 (0)	22 (71.0)
Shunt surgery	6 (12.5)	0 (0)	6 (19.4)
Liver transplant	3 (6.3)	0 (0)	3 (9.7)
Hepatopulmonary syndrome, n (%)	2 (4.2)	0 (0)	2 (6.5)
Death, n (%)	1 (2.1)	0 (0)	1 (3.2)

Table 1. Description of baseline characteristics and outcomes of patients with PSVD with and without associated portal hypertension (PHT).

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OC87

Transition of young people with chronic hepatitis B from a paediatric liver centre to adult health services

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Young people (YP) are more likely to engage and be retained if successfully linked to Adult Health Services (AHS) during the transitioning process. The aim of this study was to determine the incidence of YP with chronic Hepatitis B (CHB) successfully transitioning from a specialist paediatric viral hepatitis care to AHS and to identify any barriers that may contribute in non-linkage to AHS.

This is a retrospective study of YP followed up at a specialist paediatric liver centre in the United Kingdom who were transitioned to AHS. The study period was between 2018-2023 and includes the 2020-2021 lock down period due to SARS-CoV- 2 pandemic.

Non linkage to AHS was defined as nonattendance of at least one of the two initial outpatient appointments in AHS after transitioning. Demographics, mode of transmission, ethnicity, follow up in paediatric and adult services, antiviral treatment, first degree family member with CHB, virtual or in person consultation at transition, comorbidities, was obtained from electronic data base at paediatric and AHS sites.

Seventy YP were transitioned to AHS during the study period. Of these, 35 (50%) were transitioned during the lock down period. The mode of transmission was vertical in 44, horizontal in 4 and not known in 22. Majority were Asians (n=42, 60%).

None of the patients were lost to follow up during paediatric follow up. Of the 37 and 31 YP who were offered in person and Virtual transition appointments respectively at the paediatric centre, 23 attended in each group. 23 patients had received antivirals for CHB either as part of research study or for clinical reasons. 13 patients were on antivirals (Entecavir or Tenofovir) at time of transition.

Sixty-one YP (87%) were successfully linked to AHS while 9 did not attend AHS and were lost to follow up. 16 YP had co morbidities (other medical illness needing medical follow up) and all were linked to viral hepatitis services in AHS.

Of the 9 patients not linked to AHS, a referral to AHS was lost in the system, one YP was asylum seeker, one had high nonattendance in paediatric services, two had moved out of area and not contactable and no cause could be identified in the remaining four. The dedicated paediatric viral hepatitis service contributed to high rates of successful linkage to AHS. Of the 9 YP not linked to AHS, 7 transitioned during the lock down and may have contributed to non-linkage of care.

Table 1: Young People with CHB transitioned to adult health services

		Linked to AHS n = 61	Not linked to AHS n = 9
Transition appointment at Paediatric centre	In person	21	2
	Virtual	21	2
	By referral letter only	2	0
	During Lockdown	28	7
Family member with CHB under medical follow up•		49	6
Duration of Paediatric follow up median years(range)		8(0-20)	8(2-16)
Frequency of paediatric follow up	Biannual	17	1
	Annual	44	8
Antivirals	Past treatment with anti-viral	21	2
	Transition on anti-viral	13	0
Sex	Male	41	8
	Female	20	1

OC88

The epidemiology of paediatric metabolic-associated steatotic liver disease: a regional cohort study

OC89

Mortality risk of diabetic ketoacidosis-associated paediatric acute liver failure in the post-COVID-19 era.

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Recent reports have highlighted increasing incidence of diabetic ketoacidosis (DKA) cases, following COVID-19.[1,2] While diabetes is linked with hepatic complications, its association with paediatric acute liver failure (ALF) remains poorly understood.[3] An unusual trend of post-pandemic DKA-associated ALF, has emerged at the UK's largest paediatric hepatology centre, resulting in intensive care admission and two fatalities.

Children admitted with an initial DKA presentation to a single UK paediatric hepatology centre, in ALF, between January 2010-August 2024 were identified - all three presented after 2020. Demographic, clinical and laboratory data was collected.

Patients A (16-year-old female, BMI 20 kg/m²), B (11-year-old male, BMI 27 kg/m²), and C (11-year-old male, BMI 31 kg/m²) were identified. All three presented with DKA; A (mild; pH 7.27), B (severe; pH 6.82), and C (severe; pH 6.9), with no prior diagnosis of diabetes. Patient A presented with a peri-anal abscess, while Patients B and C exhibited reduced consciousness, requiring immediate intubation and ventilation. A head CT revealed cerebral oedema in Patient C. The national UK DKA management protocol was followed, and broad-spectrum antibiotics were initiated. On admission, liver and renal function tests were normal. DKA resolved in the following timeframes: A (6 hours), B (48 hours), and C (72 hours). However, on day 4, all three developed ALF, marked by elevated ALT levels (A: 3615U/L, B: 9984U/L, C: 4245U/L; figure 1i), peak INR (A: >15, B: 4.4, C: 3; figure 1ii), metabolic acidosis and hyperlactatemia (peak lactate A: 19.8mmol/L, B: 23mmol/L, C: 7.9mmol/L). Patient A required intubation and ventilation. ALF management protocol was initiated, including N-acetylcysteine. On day 4, Patients B and C developed cardiovascular instability and acute kidney injury, with Patient A following on day 5. Peak creatinine levels were A: 81µmol/L, B: 86µmol/L, and C: 100µmol/L (figure 1iii). All three required escalating inotropic support, haemofiltration, and urgent transfer to the paediatric hepatology centre. Prior to ALF, all patients received therapeutic paracetamol (A: 60mg/L, B: 24mg/L, C: 16mg/L). A hyperinflammatory state was evident in all patients. Microbiology investigations revealed; A - abscess; E*scherichia coli, Group B streptococcus;* B - nasopharyngeal aspirate; metapneumovirus and C - culture-negative chest infection. Viral, immunology, and metabolic investigations for ALF were negative. All three patients progressed to refractory shock. Cardiac arrest occurred on day 6 for all patients, leading to the deaths of Patients A and B. Liver autopsy in the deceased patients showed centriacinar necrosis and microvesicular steatosis. Patient C survived, with liver function tests normalising by day 64.

In the post-pandemic era, our centre has observed a severe hepatic phenotype associated with DKA, mirroring the rising incidence of DKA during this period. Factors such as ischemic hepatitis, medications[4], and sepsis likely exacerbate hepatic vulnerability in patients with pre-existing metabolic susceptibilities.[5] The increasing prevalence of obesity may further elevate this risk, contributing to our observation.[6] In addition to promoting early recognition of DKA, we urgently recommend incorporating liver function monitoring into UK DKA guidelines to facilitate early detection of liver injury and reduce associated mortality.

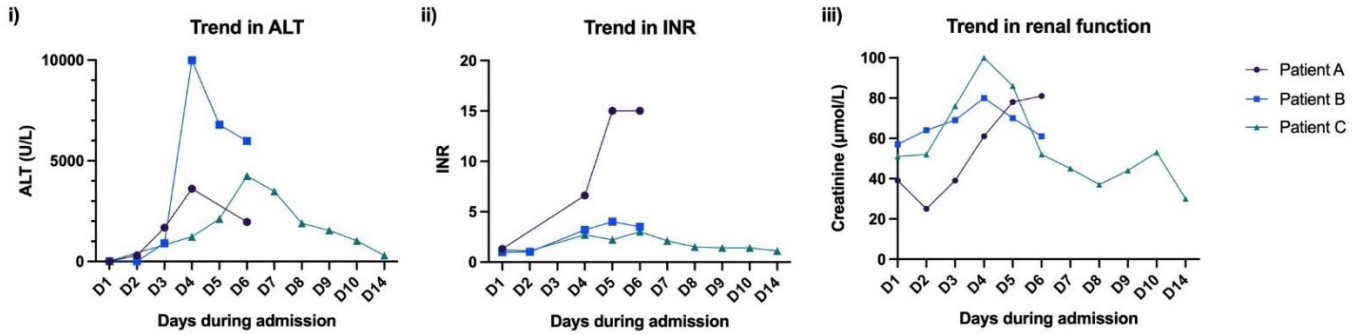

Figure 1. Trend in i) alanine aminotransferase (ALT), ii) international normalised ratio (INR) and iii) renal function using creatinine as a maker, across the admission course of Patients A-C.

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OC90

Switching fat soluble vitamin preparations in children with biliary atresia: a single-centre clinical and cost-efficient approach

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Infants with biliary atresia are typically discharged with multiple medicines following their Kasai portoenterostomy procedure. These include fat soluble vitamin supplements given whilst the child remains cholestatic and continued for the first year. A nationwide shortage of the licensed vitamin E liquid (alpha tocopheryl) in the UK, forced a change from a triple vitamin regimen consisting of a standard multivitamin preparation containing vitamin A 5000 units and vitamin D 400 units in 0.6mL (Dalivit®) plus vitamin E liquid 50mg plus phytomenadione 1mg (Konakion® ampoules) to a fat soluble vitamin combination product designed for children with cystic fibrosis containing 1600 units vitamin A, 600 units vitamin D, 40mg vitamin E and 2mg phytomenadione in 0.1mL in an MCT oil base (Paravit CF®).

The aim of this study was to assess the efficacy and cost effectiveness of the new regimen compared to the original one.

Infants with biliary atresia who underwent a Kasai portoenterostomy and cleared their jaundice with a full year of follow up on each of the vitamin regimens, from 2021 to 2024 were included. Those whose Kasai operations failed were excluded as their vitamin requirements were highly variable depending on their clinical course and timing of transplantation. Five children met the criteria in each vitamin regimen group. Vitamin levels and INR were recorded at around 6 and 12 months at which point the vitamin supplements were stopped.

The first group (triple vitamin regimen) were 2 male, 4 Caucasian, 1 Asian, all cleared their jaundice by a median of 4 months (range: 3.5-6 months) and maintained normal liver function tests. One child had an episode of cholangitis. The vitamin levels and INR were normal by 6 months in all 5 children and remained so at 12 months. Of note 3 children had vitamin D levels above the normal range with levels of 102-137nmol/L (normal range 50-100nmol/L).

The second group (single combination preparation) were 3 male, all Caucasian and cleared their jaundice by a median of 3 months (range: 1.5-5 months), although 2 had intermittent raised ALT, 1 with presumed cholangitis. The vitamin levels and INR were normal by 6 months in 4/5 children. One had a low vitamin A level which normalised by 12 months. Vitamin D levels were high in 3/5 children (104-155nmol/L).

The triple vitamin regimen has an indicative NHS cost of £150 per month per patient. The single combination preparation has a cost of £40 per month (1). The overall cost saving to the healthcare system is £1,300 for 1 year of treatment per patient.

In conclusion, the switch to a single combination product was of similar efficacy to the previous triple agent regimen and was more cost effective, saving NHS resources including consumables (only 1 oral syringe needed instead of 3) as well as reducing drug costs. It also decreases the medicine burden for parents, and this is likely to improve adherence and quality of life (2).

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OC91

Evaluating current practice of managing patients with Coeliac Disease against European (ESPGHAN) standards of care.

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An ESPGHAN position paper on follow-up of children and adolescents with Coeliac Disease (CD) (2022)[1] recommended ten standards of care. We aimed to assess the historical compliance of a busy district general hospital (DGH) service with these guidelines.

A retrospective review was completed of CD patients (N=54) in the paediatric gastroenterology clinic at a DGH from 01/01/2010 to 31/07/2023. Data were collected from health records using a specifically designed audit tool. Diagnostic criteria were assessed against the latest criteria at the time of diagnosis, and ongoing management against the ESPGHAN recommendations[2,3].

Mean age at diagnosis was 6.7 years. Serological diagnostic criteria were met in 69%, and 31% diagnosed histologically. One patient did not meet the contemporaneous diagnostic criteria but had borderline serology with persistent symptoms.

All patients received follow-up; 93% within 3-6 months of commencing a gluten-free diet; 98% every 12-24 months afterwards. All patients were followed up by a consultant, with 87% seen by a dietician.

Approximately 85% were signposted to Coeliac UK for further information and support.
All patients had serial aTTGs measured as appropriate.

Compliance to ESPGHAN's initial blood test recommendations[1] was varied. Full blood count was measured within 6 months of diagnosis in 89% of patients; liver function tests in 83%; thyroid function tests in 76%; vitamin D in 76%; iron/ferritin in 62%; and vitamin B12 in 24%.

Deficiency	% tested	% deficient	% supplemented
Iron/Ferritin	80%	56%	100%
Vitamin D	96%	83%	98%
Vitamin B12	46%	0%	0%
Folate	50%	11%	67%

Table 1: *Deficiencies investigated since diagnosis.*

All patients had gastrointestinal symptoms and adherence to a gluten-free diet assessed at each follow-up appointment, 94% had extraintestinal symptoms assessed.
Height and weight were stated in 94% and 96% of follow-up appointments, respectively, with centiles stated in 72% and 76%.

Quality of Life (QoL) was not formally assessed in any patient.

Diagnostic criteria were satisfactorily met. Fewer avoidable biopsies have been performed since the 2020 guidelines[2] were released (0%, compared to 18%), perhaps reflecting a benefit of a simpler diagnostic algorithm.

An order set of monitoring investigations has been put in place to ensure that all appropriate bloods are requested in future.

Proformas for both initial diagnosis and annual review appointments have been updated to ensure adequate compliance with ESPGHAN recommendations.

The ESPGHAN guidelines[1] recommend QoL scores for children with CD[4], plus referral onto a psychologist with CD knowledge if required. No such service exists in this DGH or region, therefore these recommendations were not met, and current service provisions will be explored further.

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OC92

An audit of the diagnostic practices for suspected coeliac disease in paediatric patients at our institution compared to the 2020 ESPGHAN guidelines

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Background

The 2020 ESPGHAN guidelines(1) on the diagnosis of coeliac disease (CD) highlight the pathway to serological and histological diagnosis. Both biopsy and non-biopsy approaches rely on the patient consuming gluten throughout investigations(2). At this institution, some individuals referred with positive coeliac antibodies had been advised to exclude dietary gluten prior to confirmed diagnosis. This could lead to reluctance in reintroducing gluten which in turn causes delays in diagnosis, inaccurate diagnosis, negative patient/family experiences and poor utilisation of specialist outpatient clinic time.

Aim

This audit aimed to compare current diagnostic and referral practice at our institution with the 2020 ESPGHAN guidelines for diagnosis of CD. The audit looked at the following objectives:

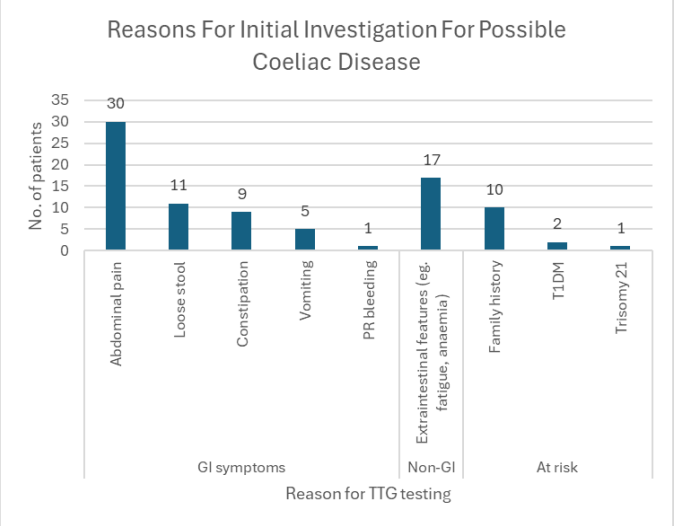
- Identify the number of referred patients that have been advised for gluten free diet (GFD) prior to formal diagnosis/exclusion of diagnosis
- Assess the duration from initial testing to confirmation or exclusion of diagnosis

Method

Patients undergoing investigation for CD between March 2023 and February 2024 were identified using a multisystem approach. Relevant data were extracted from online patient records to assess TTG levels, EMA positivity, whether on a GFD prior to diagnosis and whether they had an endoscopy. The collected data were then analysed to check if the diagnostic pathway followed the ESPGHAN guidelines.

Results

66 patients were identified for inclusion in the audit, with a mean age of 9.5yrs (range 2-17), 56% female (n=37). 65% of patients had at least one GI symptom (n=43) and the rest had no reported GI features despite having a raised initial TTG.



Over 1 in 4 patients (n=18) were incorrectly advised to exclude dietary gluten prior to full investigation for CD. Additionally, one parent had put their child on a GFD without being advised to do so, precluding full investigation for CD without first reintroducing gluten to the diet for a number of weeks. When patients had prematurely started a GFD prior to confirmed diagnosis, time taken for diagnosis/exclusion of CD was on average 4 months longer than for patients on a gluten-containing diet throughout investigation. Importantly, a sixth of those who had initially been advised to go on a GFD were later confirmed not to have CD (n=3).

62% of patients were given a diagnosis of CD, however 12% of those did not strictly follow the guidelines for diagnosis (n=5). About 1/5 of those referred with raised TTG had a diagnosis of CD excluded following comprehensive investigation, 3% were expected to have latent CD, and 15% were still awaiting investigation results at the time of data analysis.

Conclusion

This audit showed over 90% adherence to ESPGHAN guidelines. Over 60% of patients with raised TTG had CD, however 2/3 of patients required histological confirmation. Over 1/4 of patients were wrongly advised to start a GFD prior to formal diagnosis. This resulted in an average delay in diagnosis of 4 months. Following this audit, the department worked to educate local GPs, through newsletters and practical meetings, on the need to continue consuming gluten pending full investigation.

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OC93

Coeliac crisis: case report

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Coeliac crisis is a medical emergency, rarely encountered in clinical practice. It may be a consequence of a delayed diagnosis of coeliac disease (CD). Diagnostic criteria for coeliac crisis include severe dehydration including orthostatic changes, renal dysfunction, neurological dysfunction, metabolic acidosis, abnormal electrolyte levels, hypoproteinaemia with Albumin below 3g/dl and weight loss more than 4.5kg. A diagnosis is made if more than 2 criteria are present (1).

We describe the case of a 4-year-old girl who presented with coeliac crisis as the first manifestation of undiagnosed coeliac disease, to raise awareness of the diagnostic criteria for coeliac crisis and consideration of this in unwell patients.

Our patient presented with chronic diarrhoea, fatigue, and exhaustion. On admission she was not clinically dehydrated, had peripheral oedema, mildly distended abdomen with no signs of ascites. She had 20% weight loss, and her BMI was 13.23 (on 2d centile). A no-biopsy diagnosis of coeliac disease was made as she had IgA anti-tissue transglutaminase antibodies more than ten times the upper limit of normal and positive endomysium antibodies (5). Bloods showed compensated metabolic acidosis, with bicarbonate of 13, normal glucose at 4.1, low magnesium at 0.39 mmol/L, low albumin at 13 g/L. She required magnesium and albumin infusions for deficit correction. In addition, she had deranged PT/INR which normalised after two doses of intravenous Vitamin K. She also had transaminitis, and an extensive hepatitis screen. Autoimmune hepatitis was ruled out. An abdominal ultrasound showed fatty liver and normal elastography. Metabolic screen was normal. The levels of Vitamin D, folic acid, copper and zinc were low, and she had supplementation.

Our patient met 4 out of the 7 criteria for coeliac crisis. She was commenced on gluten free diet, initially via nasogastric tube, then orally from day 4. She was monitored for refeeding syndrome and discharged following nutritional rehabilitation after 11 days. At her 2 months clinic follow up her bloods were normal; she has gained weight. Four months after her presentation her BMI improved to 17.7 (on 91st centile).

Coeliac crisis was first described by Andersen and Di Sant' Agnese in 1953 when they reported 35 patients with coeliac crisis, among which 3 were complicated by fatality (3,4). It is rarely encountered, probably due to better screening and early diagnosis; however, this can occasionally be the initial presentation of CD. The only effective treatment is a life-long gluten free diet. In coeliac crisis, the emergency treatment of electrolyte abnormalities is followed by gluten free diet and monitoring for refeeding syndrome. The role of corticosteroids in coeliac crisis is controversial but there are case reports of coeliac crisis with a good response to steroids, as they can reduce intestinal inflammation and restore the brush border enzymes (2).

Our patient did not require steroids and responded well to gluten free diet. This case highlights the importance of considering coeliac crisis in an acutely unwell patient with chronic diarrhoea, weight loss and metabolic derangements.

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OC94

Obscure gastrointestinal bleeding in a child post liver transplant: a case report

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Obscure gastrointestinal (GI) bleeding may cause refractory anaemia and require repeated endoscopic evaluations. In addition, this may require iron supplementation or blood transfusion if the fall is precipitous.

GI bleeding in children with previous abdominal surgery can be associated with anastomotic ulcers and may require endoscopic intervention. We describe the case of a 10 -year-old boy, who had DBD (donation after brain death) liver transplant for cryptogenic liver disease at 2 years of age. He was subsequently diagnosed with Niemann-Pick type C at 6 years of age. He presented with multiple episodes of melena, over the course of a 7-year period, requiring numerous endoscopies to delineate the cause.

Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular disorder characterized by multiple venous malformations primarily affecting the skin and gastrointestinal (GI) tract. The lesions can affect the distal small bowel and present with either obvious melena or occult blood loss, resulting in refractory anaemia.

Pharmacological therapy includes somatostatin analogues like Octreotide in significant GI bleeding. Sirolimus (angiogenesis inhibitor) has been used as a therapeutic option with variable success.

The patient had multiple endoscopic assessments including upper GI endoscopy, ileocolonoscopy and wireless capsule endoscopy (WCE). The first endoscopic assessment was normal. A relook endoscopy identified an angioectatic lesion in the second part of duodenum to which argon plasma coagulation (APC) was applied. Double balloon enteroscopy (DBE) was planned during a precipitous episode, with a significant drop in haemoglobin of 3.5g/dl. Multiple angioectatic lesions were identified close to the Roux-en-Y anastomosis; however, haemostasis could not be achieved using clips or thermal coagulation. The patient underwent laparotomy with resection and re-fashion of Roux-en-Y anastomosis. An on the table, lap-assisted enteroscopy did not identify any other lesions. The histology of the bleeding area of the resected segment showed multiple dilated vascular channels in the submucosa, appearing to be venous in nature. There has been no further bleeding since the operative intervention in a 2 year follow up.

There are no guidelines for the management of BRBNS with GI involvement. The treatment is determined by the severity of the disease and the extent of intestinal involvement. Therapeutic options include interventional endoscopy, surgery, and angiogenesis inhibitors.

Successful treatment is reported in 92% of cases. Surgical intervention is curative once the disease site is identified. Disease recurrence is known in 25% of treated patients (2). Sirolimus has proven to be effective as a second line of treatment, but there are possible limiting side effects (bone marrow suppression with increased risk of infections, toxicity, vascular complications, hyperlipidaemia, high blood pressure, abnormal renal function, impaired wound healing, mouth ulcers, rashes, arthralgia, diabetes, malignancy).

Our case study suggests that the diagnosis of BRBNS lesions can be challenging, especially in children with previous abdominal surgery and without any cutaneous stigmata. DBE during an acute episode can be useful to localise the lesion and treat it using haemostatic clips, injections, diathermy probes. Surgical help maybe needed in more extensive lesions or when endoscopic intervention is difficult or not possible.

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OC95

Audit of practices for patients with Coeliac Disease in peripheral gastroenterology clinics.

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Coeliac disease is an autoimmune condition, damaging the gut due to exposure to gluten in food. Non-compliance leads to long term health complications.
We wanted to audit our practices against the National Institute for Health and Care Excellence clinical guideline (NICE CG20).

We identified presenting age, symptoms and nutritional deficiencies. We looked at the compliance as recommended by NICE: annual review, surveillance bloods, information given about immunisation, long-term health effects from non-compliance and dietary management.

NICE CG20 and ESPGHAN coeliac disease guidelines were used as the standard to audit our practice.

Retrospective data was collected over 12 months from paediatric gastroenterology clinics only, for all patients with diagnosis of coeliac disease using a proforma. The sample size was 54 patients.

Analysed data showed:

1. Age of presentation: 29% >12 year, 46% 6-12 years and 24% 1-5 years old

2. 74% had GI symptoms, 26% had tiredness and 11% had dizziness

3. 85% diagnosed serologically within this cohort.

4. 48% had ferritin and vitamin D deficiency and 44% had microcytic hypochromic anaemia at diagnosis.

5. 83% had annual review, 94% had surveillance bloods and discussion about long-term health effects.

6. 46% only had documented discussion about immunisation and 87% were signposted to Coeliac UK resource.

7. 100% had dietetic referral, 94% had dietetic review and discussion about nutritional deficiencies.

We showed:

1. We are following NICE coeliac guidelines for monitoring, information support and dietetic review. We are following ESPGHAN coeliac disease guideline for diagnosis.

2. Difficulty is when a patient is referred from out of the area (therefore not having follow up by our area dietician) or are non-compliant.

3. Need to standardise the monitoring and the information given in the outpatient clinic via the use of a
   - Coeliac disease checklist (below)
   - Use of information leaflet (not attached)

NHS

NHS Trust

Surname	Unit No
Forename	NHS No
Address	DOB
Postcode	(or affix patient label)

Coeliac Disease Checklist

Date:.....

Time: .....

Clinic:.....

Growth centile:.....

Height: ..... Height centile: .....

Weight: ..... Weight centile: .....

Confirm Diagnosis:

Serology / Biopsy date:.....

Explain diagnosis of coeliac disease:

- ☐ Coeliac disease is a lifelong autoimmune disease
- ☐ The only treatment is a strict gluten free diet
- ☐ Even tiny amounts of gluten can cause symptoms and gut damage
- ☐ Awareness about nutritional deficiencies
- ☐ Surveillance bloods ☐ Bloods at diagnosis

☐ Gluten free diet ☐ Not yet ☐ Already on it

Resolution of symptoms: ☐ N/A ☐ Some ☐ All

☐ Avoid cross contamination

☐ Following ☐ Not following UK immunisation programme

☐ Signposting to resources: Coeliac UK website

☐ Non-compliance can lead to serious health issues: brittles bones, intestinal lymphoma, etc.

☐ Flour and bread on NHS prescription in primary care where applicable

☐ Screening first degree relatives

Dietetic referral - Referred: ☐ NXH ☐ Outside area ☐ already under dietetics

Name:..... Signature: .....Designation: .....

Date:.....Time: ..... Stamp

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OC96

Childhood Stroke and Coeliac Disease – A Case Study

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Childhood stroke is a rare occurrence, with around 400 children in the UK experiencing a stroke each year. Approximately one-third of childhood strokes are cryptogenic, meaning that the underlying cause remains unknown [1,2]. An association between coeliac disease (CD) and acute ischaemic stroke has been suggested due to the influence of chronic inflammation, abnormal immune response, endothelial damage and prothrombotic states [2-9]. Here, we present a case of stroke in a 4-year-old girl with no known medical history who was later discovered to have CD.

A previously healthy 4-year-old girl presented to the emergency department (ED) following a syncopal episode along with new left sided weakness and a one-day history of right sided headache. Her neurological assessment revealed unilateral weakness on the left side with reduced ability to hold her limbs against gravity and fatigue when walking without support. Her initial bloods revealed severe anaemia with Haemoglobin 4.8 g/dL, other blood counts were normal, including normal clotting profile. CT head revealed a thrombus in the entire internal cerebral vein. Extensive further bloodwork, including iron studies, haemoglobinopathy screen, thrombophilia screen and coeliac screen, confirmed iron deficiency anaemia and CD with positive anti-tissue transglutaminase IgA (titres >128) and IgG (titre 69.0), and positive IgA endomysial antibodies. This resulted in a non-biopsy diagnosis of CD.

She received fluid boluses, one blood transfusion, and was commenced on treatment dose low-molecular-weight-heparin (LMWH) 100 units/kg twice daily (BD) following discussion with the tertiary paediatric haematology team. Conservative treatment with 72-hours of hourly neuro-observation and monitoring for signs of raised intracranial pressure was carried out following discussion with tertiary paediatric neurology and neurosurgical teams.

Following 72-hours of observation, she was discharged with 5-days LMWH 100 units/kg BD then switched to oral rivaroxaban 5mg BD for 3-months. Her repeat MRI showed that the deep cerebral veins, Vein of Galen and Straight sinus had recannalised. She is currently doing well, is asymptomatic and on a lifelong gluten free diet with no further acute ischaemic stroke events reported.

Some studies have reported that CD is associated with an increased risk of cardiovascular disease, due to its association with other autoimmune diseases including autoimmune vasculitis, which can directly affect the cerebral vessels and increase the risk of stroke [4,7,9]. This combination of chronic inflammation, cross-reacting antibodies and endothelial dysfunction may influence the higher incidence of ischaemic stroke, myocardial infarction and thromboembolism seen in those with CD and suggests that there is an autoimmune component for CD associated vasculopathy [7,9].
We suggest that CD is a potentially treatable cause of cerebral vasculopathy, and CD serology should be considered when investigating cryptogenic stroke in childhood, even in the absence of typical gastrointestinal symptoms. It highlights the importance of considering CD as a differential diagnosis for ischaemic stroke, allowing for earlier diagnosis and management of CD.

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OC97

Attitudes Towards Ergonomics in Endoscopy: Results of a BSPGHAN Endoscopy Working Group National Survey

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Ergonomics, in the context of endoscopy, refers to the study and optimisation of an endoscopist's interaction with their endoscopy environment, to reduce the rate of endoscopy-related injury (ERI).(1) While the risk and prevalence of ERI has been established in adult gastroenterology,(2–4) and paediatric gastroenterologists in the USA,(5) there are no available data on this subject amongst UK paediatric gastroenterologists. Considering increasing awareness of this issue, the American Society for Gastrointestinal Endoscopy developed guidelines for prevention of ERI, and a core curriculum presenting best practice recommendations for ergonomics in endoscopy, highlighting the importance of optimisation of factors such as posture, room set up, and appropriate equipment handling in paediatric endoscopy.(1,6) However, ESGE/ESPGHAN joint guidelines on paediatric endoscopy do not mention ergonomics and there exists no similar guidance for the European audience.(7)

Aim: To ascertain attitudes towards ergonomics in endoscopy and the prevalence, nature, and risk factors for endoscopy-related injury in the UK paediatric endoscopist population.

Methods: A 26-point questionnaire survey focusing on three sections: demographics and clinical experience in endoscopy; history of injury related to endoscopy; previous experience with ergonomic modification in endoscopy was distributed to all members of BSPGHAN via email and social media.

Results: 65 respondents. 50.8% (n=33) female. 52.3% (n=34) aged 35-44 years, 24.6% (n=16) aged 45-54 years). 53.8% (n=35) consultant grade, the remainder were registrar, clinical fellow, or SAS. 76.9% (n=50) performed 1-4 hours of endoscopy weekly, 18.5% (n=12) practiced 5-10 hours weekly. 53.8% (n=35) stated they had previously experienced injury or pain related to endoscopy. Of those who had experienced injury/pain, the most common experiences were hand/digit pain (62.2%, n=23); backache (35.1%, n=13); arm pain (24.3%, n=9); neck ache (16.2%, n=6). 29.2% (n=19) had experienced pain during endoscopy monthly, while only 18.5% (n=12) had taken analgesia for endoscopy related pain. 86.2% (n=56) had never had any training focused on ergonomics and/or reducing ERI. Only 6.2% (n=4) reported that ergonomics formed part of their theatre brief. There were significant differences in rates of endoscopy-related injury in females vs males (69.7% vs 37.5%, p=0.013), as well as in respondents who reported performing weight/resistance exercises vs not (39.4% vs 68.8%, p=0.025). There was no significant difference between rates of injury between consultants vs non-consultants (54.3% vs 55.5%); those using water-assisted colonoscopy vs not (36.9% vs 60.1%, p=0.103); age <45 years vs >45 years (56.1% vs 50%, p=0.79); and those performing cardiovascular exercise vs not (63.6% vs 51.9%, p=0.52).

Conclusion: This is the first UK nationwide study of ergonomics and ERI amongst paediatric endoscopists. Over half of paediatric endoscopists surveyed experienced ERI, most commonly hand/digit pain, backache, arm pain, and neckache. The vast majority of endoscopists had never had training in ergonomics. We found that women and endoscopists not performing resistance exercises were significantly more likely to experience ERI. These findings provide impetus for the development and delivery of training and guidelines in ergonomics to reduce the risk of occupational injury amongst UK-based paediatric endoscopists.

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OC98

Host-Microbiome and Immune-Related Protein Interactions in the Preterm Milk Fortification in Neonates (PUFFIN) Study

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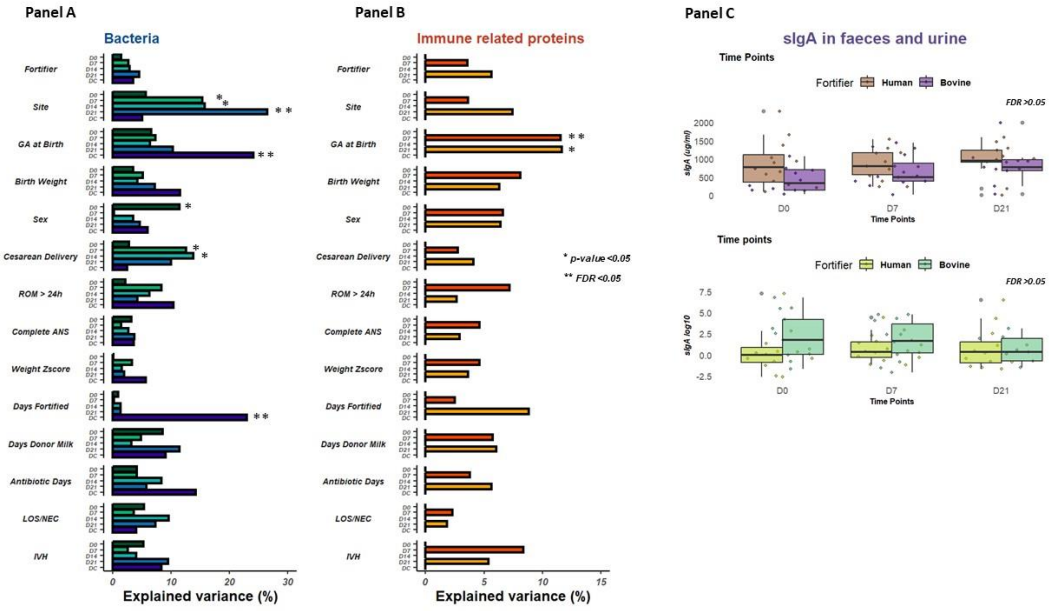
Most infants born before 32 weeks gestation will need fortification to meet nutritional requirements (1,2). Fortifiers can be bovine milk-based (BMF, usually powdered) or human milk-based (HMF, previously liquid) and uncertainties remain about potential benefits of HMF (3). Powdered HMF preserves maternal milk volume in the infant diet and may offer greater immunomodulatory and less inflammatory benefits compared to BMF.

Within a randomised clinical trial of powdered BMF (PBMF) vs powdered HMF (PHMF) in 28 infants <32weeks gestation otherwise receiving only human milk we compared: faecal microbiota, stool and urinary secretory immunoglobulin A (sIgA), and stool cytokines from first fortification (commenced at 150mls/kg/day of milk feeds) until discharge. 129 stool and 109 urine samples were analysed from infants of median gestation 27 weeks. Faecal samples were analysed for sIgA (ELISA), 39 immune proteins (MSD V-PLEX), and microbiome (16S rRNA sequencing), while urine samples were assessed for sIgA (ELISA). While stool sIgA levels appeared higher in the intervention group at day 21, the difference was not statistically significant. However, when assessing sIgA progression across all time points until discharge, levels were significantly higher in the PHMF group (p = 0.04). The fortifier type did not directly impact other measures.

Gestational age at birth and unit of care (two recruited units) had a significant impact on microbiome and cytokine profile One NICU using *Bifidobacterium*-based probiotics had higher relative abundance of this genera (p = <0.05) and distinct immune related proteins profiles.

Multi-omic analysis revealed a positive correlation between sIgA and Enterobacteriaceae (p = <0.05), with additional correlations among *Staphylococcus*, *Bifidobacterium*, *Escherichia*, and specific immune proteins. No significant differences in microbiota composition or cytokine profiles were found between PHMF and PBMF, unlike previous studies with liquid fortifiers (4). PHMF appeared to increase stool sIgA levels longitudinally until discharge, but no significant differences were detected at individual time points. The clinical significance of this finding remains uncertain. Covariates, including gestational age and routine probiotic use play a role in shaping microbiota composition and cytokine profiles, potentially masking smaller impacts of fortifier type. These findings highlight the complex interactions between clinical variables, gut-health, microbiome, and immune responses in preterm neonates. Further research in larger cohorts is needed to evaluate the clinical implications, efficacy, and cost-effectiveness of fortifiers.

Figure 1.



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OC99

Prevalence of Zinc and Copper Deficiency among children receiving jejunal nutrition.

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Aim of study:
The objectives of this study are to review the prevalence of zinc and copper deficiencies among children who have been on jejunal nutrition.

Background:
In jejunal fed children, we found high prevalence of micronutrient deficiency and often struggled to find proper guidelines to follow to manage and prevent deficiencies.

Methods:
A retrospective review of notes, electronic record of patients, dietic notes and blood levels of Zinc and Copper in children who have been on jejunal nutrition between 2018 to 2024.

Results:
There were 86 children on jejunal nutrition between 2018-2024, out of which 40 children had supplemental oral nutrition. Out of 86 children, 38 children are currently continuing on JEJ feeding, 26 children have transitioned to other types of enteral feeding. 17 children passed away while on jejunal feeding and the feeding status of 5 children is unknown.
The most common age jejunal nutrition was started is < 1 year, accounting for 27 in number. The average age of starting jejunal feed is 5.2 years and the youngest is 3 weeks old. The average duration of jejunal feeding is 3.3 years.
In our cohort, prior to the year 2022, the average micronutrient monitoring from the year 2018 to 2021 is seen at 48.75%. Micronutrient monitoring after 2022 improved seen after the appointment of nutritional nurses in the department, averaging at 68% in 2022, 80% in 2023, 61.5% by mid-2024.
In the study group, 28% of children were found to be deficient in zinc and 17% of children were deficient in copper. Of those deficient, 33% had low albumin and 50% had high CRP at the time of zinc level measurement. Overall, in this study, it was seen that after starting jejunal nutrition , micronutrient deficiencies in copper and zinc and selenium were significantly reduced.

Conclusions:
This study done on the pediatric population showed that micronutrient deficiencies like copper and zinc were significantly reduced over the course of jejunal nutrition. However, regulated monitoring of micronutrient will lead to better outcome. In our cohort, few micronutrient studies were done while children were admitted in the hospital for an illness.The incidental finding of altered low levels of these micronutrients when done in the acute phase of inflammation demands robust guidelines to regularly monitor the micronutrients as well as guidelines on early supplementation of zinc and copper in such children.

Table *1* The number of Copper and Zinc deficiencies

Year	Number of Copper deficiency	Percent	Number of Zinc Deficiency	Percent	Number of children on jejunal nutrition
2024	2	5%	2	5%	38
2023	7	15%	7	15%	47
2022	2	4%	10	18%	57
2021	3	7%	6	13%	46
2020	1	3%	5	14%	35
2019	1	4%	3	12%	26
2018	0	0%	1	2%	17
Total	16	32%	32	64%	

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OC100

Multiple micronutrient and trace element deficiencies (including scurvy) presenting in a child with Kwashiorkor from a significantly restricted oral diet.

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Adequate intakes of both macro and micronutrients are essential for the optimisation of growth and health in children. Increasing presentations of increasingly restricted diets are fuelling concerns of nutritional adequacy. When possible nutritional deficiencies are suspected ESPGHAN recommends a careful multimodal approach that includes biochemical markers, clinical examination and dietary assessment ([1]).

Aim: to describe a challenging case of multiple nutritional deficiencies.

A 10-year-old female was admitted to her local hospital with lower limb oedema and purpuric skin rash. Initial blood test showed low albumin and platelets. Background medical history included chromosome 15q1.11.2 microdeletion with specific associated symptoms of seizures, autism, epilepsy, significant behavioural issues and learning difficulties.

Growth history revealed weight tracking >75th centile and height tracking 0.4th -2nd centile for the previous 2 years. Nutritional review revealed poor progress at weaning. Diet diversity was poor and reduced further in recent years (<5 foods and carbohydrate predominant).

On admission her weight was 39.9kg (oedematous) 84th centile (z score 0.9), height 122cm <0.4th centile (z score -2.7). Mid parenteral centile was 25-50th. BMI was 26.8 >99th centile (z score 2.7).

The patient was transferred to a tertiary centre with concerns of Kwashiorkor.

Scurvy was initially suspected. Biochemical markers confirmed a significantly deficient level (3.5umol/l - reference range 8.8-124umol/l). Subsequent full nutrition blood screening was undertaken. No inflammatory response was identified that may have impacted on validity. Results revealed a range of additional significant micronutrient deficiencies including zinc, selenium, copper and vitamin A.

To correctly interpret these results, dietary analysis (DietPlan6) and a nutritional focused clinical examination was undertaken. This indicated a protein intake of 4g/day equivalent to 13% reference nutrient intake (RNI). Results further confirmed multiple micronutrient intakes below the lower reference nutrient intake (LRNI). Nutritional based clinical examination ([2,3]) uncovered multiple physical signs of micronutrient deficiencies further corresponding to the levels and intakes identified (Table 1).

Nutritional treatment was initiated. Within 1 week symptoms of oedema, bruising, rash, energy levels and engagement were resolving. Repeat vitamin C showed a normal level. A plan for PEG placement was made as a longer-term solution to optimise diet.

Characteristics of chromosome 15q1.11.2 microdeletion seizure disorder have been described ([4]) however significant nutritional and growth impact is an unreported consequence of this rare mutation.
This case highlights:
1) a high BMI, that may have been falsely reassuring to multiple professionals, was concealing both stunting and significant macro and micro nutritional deficiencies
2) highly restrictive diets can result in serious significant clinical deficiencies.

Table 1- Multimodal assessment of nutritional status

Nutrient	**Intake assessment**		**Biochemical assessment**		**Clinical assessment**
	% RNI	% LRNI	Result	Range	Observed symptom
Protein	13	*	24 (albumin)	35-52	Oedema, distended abdomen, low appetite, irritability, fatigue, stunted growth
Vitamin C	12	45	3.5	8.8 - 124	Purpuric skin rash, diarrhoea, bulging eyes, dry hair, fatigue
Copper	10	*	5.0	11.2 - 23.7	Cytopenia
Vitamin A	0	0	0.19	0.90 - 1.7	Vision issues
Zinc	3	4	6.0	9.8 - 19	Stunted growth, diarrhoea
Selenium	0	0	0.2	0.6 - 1.29	Thyroid dysfunction
Vitamin B12	0	0	1025	197 - 771	Thrombocytopenia

* N/A

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OC101

Improving Biochemical Monitoring and Management of Biochemical Derangements in Neonates on Parenteral Nutrition: A Two-Cycle Audit at a Tertiary Neonatal Unit in the United Kingdom

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Infancy is a critical period for growth and development, necessitating optimized nutrition to meet caloric requirements of 110–135 kcal/kg/day. Neonates unable to tolerate adequate enteral nutrition require total parenteral nutrition (TPN), which provides all nutrition intravenously, often alongside enteral feeds. Blood monitoring is essential for early detection of abnormalities, yet current practices vary significantly in terms of gestational age criteria for TPN, timing of initiation, frequency of biochemical monitoring, and management of derangements.

This study presents a two-cycle audit conducted in a Level 3 neonatal intensive care unit, focusing on premature infants born at <32+6 weeks of gestation who received parenteral nutrition. Data collection for Cycle 1 occurred from September to November In 2022 and 2023, assessing monitoring frequency and biochemical derangements. Based on identified gaps, we introduced a guideline specifically addressing hypertriglyceridemia, accompanied by a pre-configured order set to standardize monitoring practices.

Cycle 2 data were collected from January to March 2024 to evaluate the impact of these interventions.

The second cycle demonstrated a median initiation time for monitoring improved from 21 hours to 12 hours, with an increase in TPN initiation for neonates under 24 hours of age. However, early monitoring was not sustained due to challenges such as the transition to new order communication systems and a lack of clinical engagement. A follow-up survey was conducted to investigate these barriers further.

This audit highlights the necessity for continued education and systemic support to enhance biochemical monitoring and management practices for neonates on TPN, aiming to align with NICE/BAPM standards and ultimately improve patient outcomes.

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Muna Barakat .Treatment-related problems in neonates receiving parenteral nutrition: risk factors and implications for practice 2024

OC102

Clinical outcomes of infants born with exomphalos: 20-year experience from a UK regional institution

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Exomphalos occurs in 1/13000 live births, and is a central abdominal wall defect, allowing herniation of abdominal viscera, covered by a membrane, into the umbilical cord. Exomphalos can be defined as major or minor dependent on defect size and liver herniation (1),and is associated with comorbidities in up to 72% of cases (2). Literature on long term respiratory and nutritional outcomes associated with exomphalos is limited, compared to gastroschisis, another abdominal wall defect (3,4,5).

This study aimed to examine the longterm nutritional and respiratory outcomes in a single centre cohort of infants with exomphalos.

A retrospective cohort study of respiratory and nutritional outcomes of babies born with exomphalos in a single tertiary neonatal unit between 2000 – 2020 was carried out. Babies were identified from the clinical coding database and all infants with an abdominal wall defect were identified. Babies with gastroschisis were excluded. Case notes were reviewed and data collected on demographic details, other comorbidities, surgical management, nutritional and respiratory outcomes and mortality. For comparative purposes cases were divided into exomphalos major and minor. Relative risk (RR) with 95% confidence intervals and p values were calculated with an alpha of <0.05 considered significantly different.

Gender of infants and median gestational age at delivery was similar between groups. Preterm births are similar in both groups with a higher incidence (40% vs 3%) of postnatal diagnosis in infants with examphalos minor. 48% of infants with exomphalos minor and 75% with major had other

comorbidities. Median length of stay was 6 days in minor versus 43 days in major. Mean length of invasive ventilation was 4 days in minor compared to 13 days in major. 1(3%) infant with major required home oxygen, and 5 (14%) infants with major required tracheostomies. 1(4%) infant with minor required parenteral nutrition (PN) compared to 10 (28%) with major. Median days to reach full feeds was 4 days for infants with minor and 8 days for infants with major. All surviving infants with minor were orally fed at age 3 compared to 67% of children with major. Duration of parenteral nutrition (PN) was longer in major compared to minor. No children required home PN.

Infants with exomphalos major had longer hospital stays and higher rates of comorbidities and complications. Exomphalos minor was not associated with long term respiratory or nutritional consequences. 5 (14%) children with exomphalos major required tracheostomy, and 8 (45%) required long term enteral feeding support – all had significant comorbidities. No children required home PN.

This data offers insight into likely long term outcomes for infants with exomphalos. This is useful for developing departmental practices and for parents as part of the antenatal or postnatal counselling process, but further research as part of multi-centre networks is needed.

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